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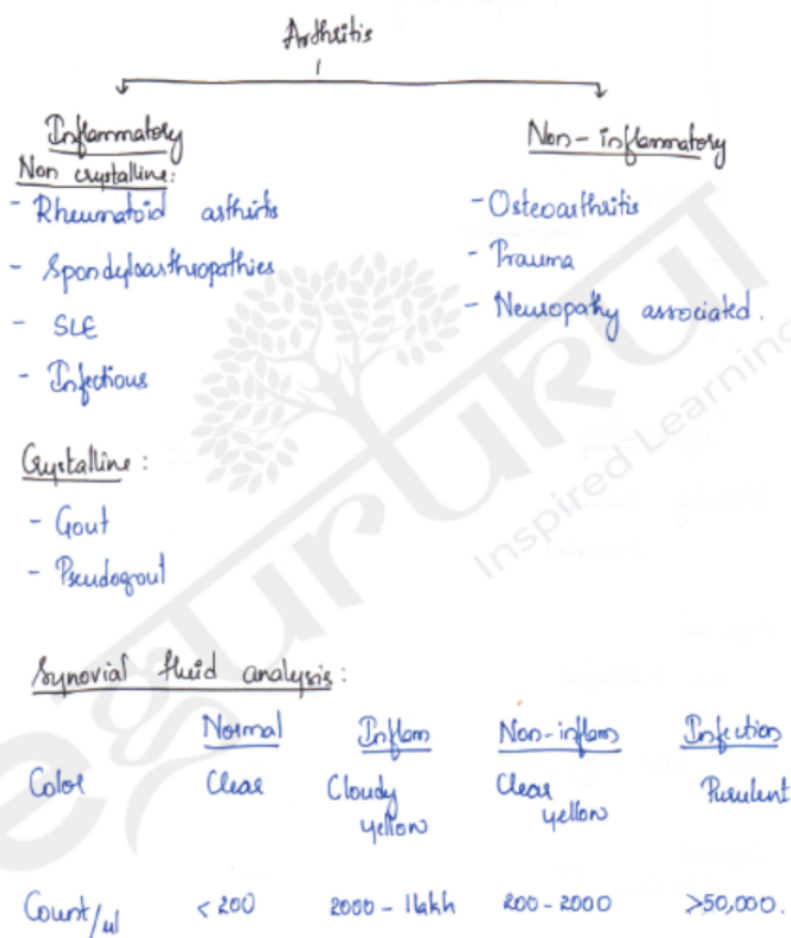
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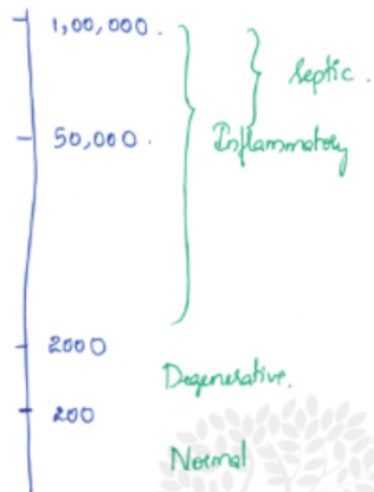
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## RHEUMATOLOGY - PART-1 ARTHRITIS

Arthritis → Inflammation of joint







### Inflammatory

- Morning stiffness  
    > 30 min
- Synovial  
    PLC > 2000/uL
- ESR, CRP - (↑)
- Systemic manifestations ++

### Degenerative

- < 30 min
- PLC < 2000/uL
- (N)
-

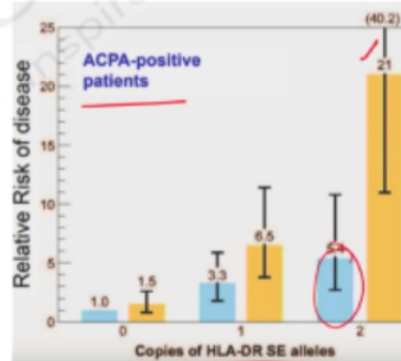
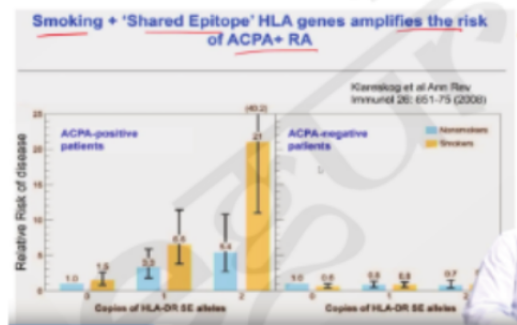
## Rheumatoid arthritis

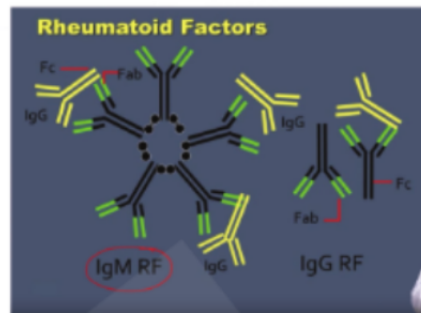
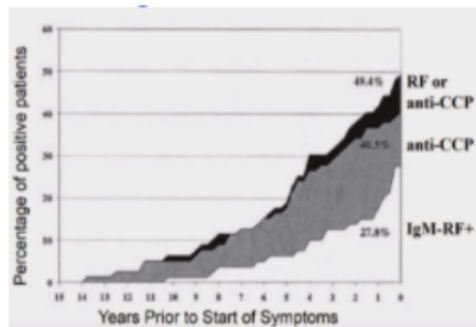
F:M → 2-3:1

HLA - DRB<sub>1</sub>



Smoking + MHC risk → Autoimmunity to citrullinated proteins





Auto Abs precede  
onset of symptoms

Rheumatoid factor  
↳ IgM against Fc of IgG  
Sn & Sp is low.

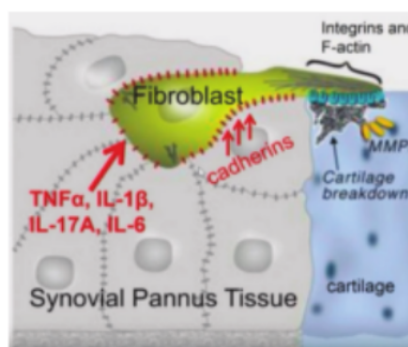
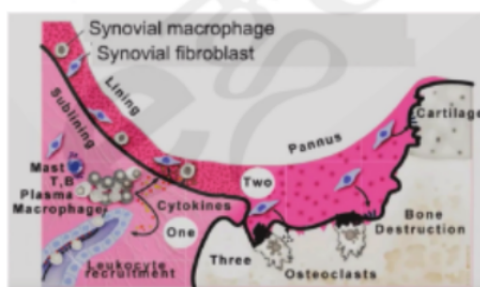
Anti CCP Abs - cyclic citrullinated peptide Abs

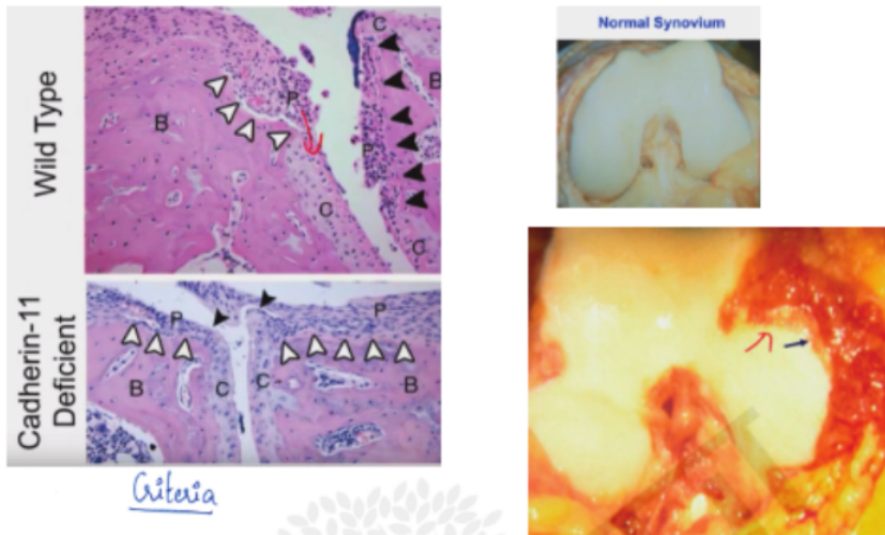
Anti MCV Abs - mutated citrullinated vimentin Abs } High specificity.

Patho:

Capillary proliferation → Pannus ⊕.

Migration of fibroblasts → erosions of bone.





### Criteria

ACR / EULAR → classification criteria

- Joint distribution
- Serology - RF  
Anti CCP
- Symptoms duration [ < 6wks  
> 6wks
- Acute phase reactants [ ESR  
CRP

Total - 10

Likely RA →  $\geq 6$

| JOINT DISTRIBUTION (0-5)                      |   |
|-----------------------------------------------|---|
| 1 large joint                                 | 0 |
| 2-10 large joints                             | 1 |
| 1-3 small joints (large joints not counted)   | 2 |
| 4-10 small joints (large joints not counted)  | 3 |
| >10 joints (at least one small joint)         | 5 |
| SEROLOGY (0-3)                                |   |
| Negative RF <u>AND</u> negative ACPA          | 0 |
| Low positive RF <u>OR</u> low positive ACPA   | 2 |
| High positive RF <u>OR</u> high positive ACPA | 3 |
| SYMPTOM DURATION (0-1)                        |   |
| <6 weeks                                      | 0 |
| ≥6 weeks                                      | 1 |
| ACUTE PHASE REACTANTS (0-1)                   |   |
| Normal CRP <u>AND</u> normal ESR              | 0 |
| Abnormal CRP <u>OR</u> abnormal ESR           | 1 |

Joints affected :  
 - MCP  
 - PIP  
 - Wrist.

DIP involved :

- |                         |                |
|-------------------------|----------------|
| - Osteoarthritis        | - Degenerative |
| - Psoriatic arthritis   | } Inflammatory |
| - Reiter's              |                |
| - Post viral arthritis. |                |

CF of RA :

- B/L, symmetric, small joint involvement  
[Swelling, warmth]
- Morning stiffness >1hr
- Fever, wt loss, fatigue.

O/E :

- DI appearing
- Ulnar deviation
- Deformity.
- Limited movement
- Extra articular manifestation:
  - o Eye : (m.c) kerato conjunctivitis sicca.
    - Episcleritis - No pain
    - Scleritis - Pain +
  - X uveitis



• Lung:

- Pleural - Pleuritis
  - Pleural effusions
    - ↳ Pl. fluid → very low glucose  
( $<30\text{mg/dl}$ )
  - Interstitial lung ds
  - Caplan's syndrome
    - ↓
    - Pulm nodules + Pneumocystis

• Heart: - Pericarditis

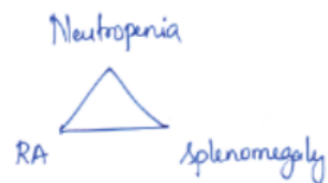
- Restrictive cardiomyopathy
- Myocarditis
- MR.
- ↑ risk of MI.

• Blood: - Anemia - Normocytic normochromic

- Neutropenia
- Lymphoma → Diffuse large B cell lymphoma  
[DLBCL]

• GI - Splenomegaly.

Felty's syndrome / triad :



- Renal : - Membranous nephropathy
  - Secondary amyloidosis
  - Nephropathy due to drugs.

★ CNS is not affected.

- PNS - - Compressive neuropathy
  - Atlantoaxial subluxation → Spinal cord compression
  - Mononeuritis multiplex.

- Skin - Purpura
  - Vasculitic rash

- Hypogonadism

• Serology :

|          | <u>Sn</u> | <u>Sp</u> |
|----------|-----------|-----------|
| RF       | 60-80%.   | 80%.      |
| Anti CPA | 40-60%.   | >95%.     |
| Anti mCV | 40-60%.   | >95%.     |

Combination  $\rightarrow \uparrow$  Sn.

Xray :

|                      | <u>RA</u>                                    |
|----------------------|----------------------------------------------|
| A - Alignment        | - Ultra deviation                            |
| B - Bone density     | - Osteopenia.                                |
| C - Cartilage spaces | - Erosions - rat bite, cysts, cartilage loss |
| D - Distribution     | - Symmetrical                                |
| E - Soft tissue      | - Functum swelling                           |

### Deformities :

- Swan neck deformity  $\rightarrow$  PIP - extension  
DIP - flexion
- Boutonniere deformity - DIP - extension  
PIP - flexion

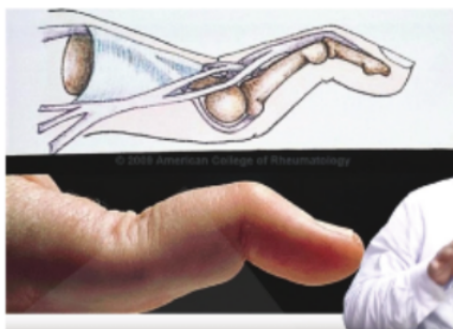


A- ulnar deviation  
B Osteopenia  
C-  
-bare area erosion  
- "rat bite" erosions  
- cysts vs erosions  
- diffuse cartilage loss  
D- symmetrical  
S- Fusiform swelling

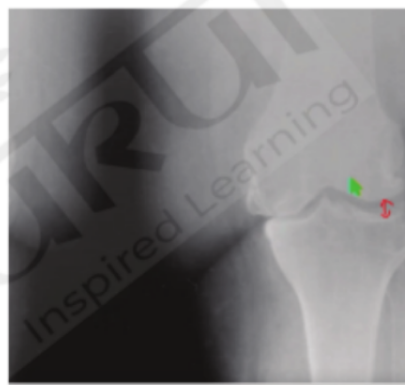
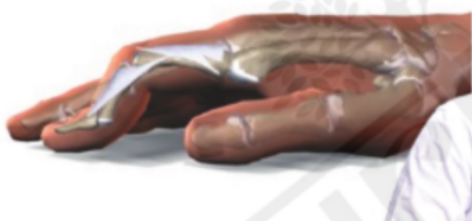


Image:





Boutonniere  
Deformity



Bouchard's nodes - PIP

Heberden's nodes - DIP

### Osteoarthritis

#### Xray

- A - Thumb deformities
- B - Normal
- C - Asymmetric narrowing  
Osteophytes - classical.
- D - Thumb base, DIPs
- E - Over osteophytes



"Gull wing" → Erosive OA.

but no mouse ear appearance  
↓  
seen in Psoriatic.

Features suggestive secondary OA:

- Unusual distribution - MCP - hemochromatosis.
- Uniform cartilage narrowing
- Large / numerous cysts
- Marked swelling.



New bone formation  
in DIP → OA.



Gull wing appearance

Unequal involvement  
of joints



Osteophyte formation.



Neuropathic joint → 4Ds - Dislocation  
Destruction  
Debris  
No demineralisation  
[No osteoporosis]

Rx of OA:

- Oral NSAIDs
- Topical NSAIDs
- Tramadol
- Intra-articular corticosteroid injections

Do not use:

- Chondroitin sulfate
- Glucosamine
- Topical capsaicin.



**Table 4. Pharmacologic recommendations for the initial management of knee OA \***

We conditionally recommend that patients with knee OA should use one of the following:

- Acetaminophen
- Oral NSAIDs
- Topical NSAIDs
- Tramadol
- Intraarticular corticosteroid injections

We conditionally recommend that patients with knee OA should not use the following:

- Chondroitin sulfate
- Glucosamine
- Topical capsaicin

We have no recommendations regarding the use of intraarticular hyaluronates, duloxetine, and opioid analgesics

\* No strong recommendations were made for the initial pharmacologic management of knee osteoarthritis (OA). For patients who have an inadequate response to initial pharmacologic management, please see the Results for alternative strategies. NSAIDs = non-steroidal antiinflammatory drugs.

## RHEUMATOLOGY - PART 2 - ARTHRITIS

\* DAS score → RA

Dr of RA:

- Steroids

- DMARDs

Hydroxychloroquine → Retinopathy.

Methotrexate } → **Anchoring drug.** - oral/s.c. - 7.5mg/wk

Leflunomide } **teratogenic** ↓ 25mg/wk.

Sulphasalazine

- Mtx → follow up with - TC, DLC, LFT. - every 3 months
- Advice against alcohol - as may damage liver.
- Leflunomide - Dihydro diolase dehydrogenase inhibitor.
  - Pregnancy should be avoided 2-3 wks after stopping the drug.
  - **Cholestyramine** - used to break enterohepatic circulation → ↑ clearance
- Sulphasalazine - TC, DLC
  - Oligospermia

### Anti-TNF drugs:

- Etanercept - Recombinant sTNF receptor
- Adalimumab - Human mAb
- Golimumab - " "
- Certolizumab pegol - Pegylated Fab fragment from humanised mAb
- Infliximab - chimeric murine human mAb.

### Triple therapy:

Methotrexate + Sulphasalazine + HCQ.

Anakinra - IL-1 antagonist

Toclizumab } IL-6 "  
Sarilumab }

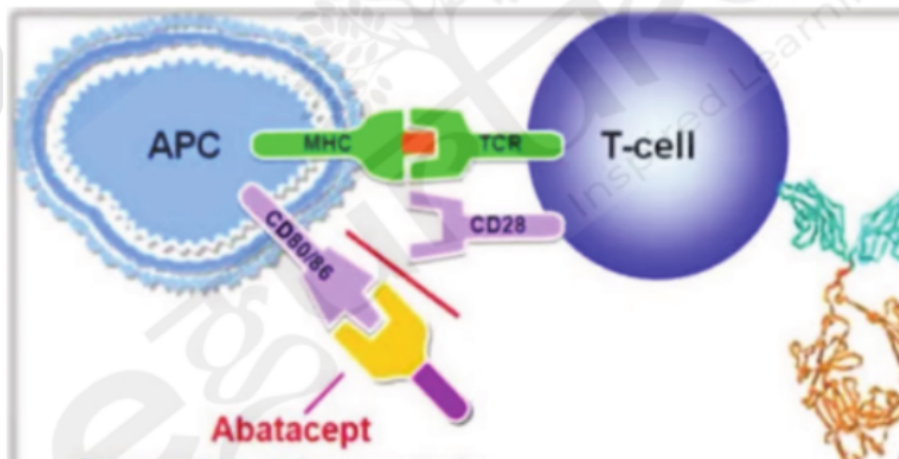
Abatacept - CTLA<sub>4</sub> inhibitor - block co-stimulation of T cells

Tofacitinib - JAK inhibitor → ↓ inflam cytokines

### Complications of Anti-TNF drugs:

- Reactivation of TB
- Neuropathy.
- Lupus like rashes
- ↑ infections.
- Cancer risk?

Avoid in - SLE  
- Hep B



## Low backache

### Inflammatory

- Pain - morning ⊕.
- Morning stiffness
- Night pain  
↳ ⊕ on walking around.
- NSAIDs - ↓ pain

Radio:

Sacro ilitis  
Syndesmophytes  
Spinal ankylosis

### Degenerative (OA)/ Mechanical

- Pain - more as day progresses  
⊖.
- Pain on exertion
- minimal response.

Osteophytes

Disc space narrowing

Vertebral malalignment.

### Spondyloarthropathy

- Ankylosing spondylitis
- Reiter's (Reactive arthritis)
- Psoriatic arthritis
- IBD associated arthritis

- M:F = 3:1
- Sacroilitis
- HLA B27
- Uveitis
- Enthesitis - inflammation of ligaments  
Achilles tendinitis  
Anterior longitudinal ligament.

- Good response to NSAIDs
- Family history (+)
- Dactylitis
- Rheumatoid factors (-) - seronegative spondyloarthritis.

### Axial Spondyloarthritis

#### 2009 ASAS classification criteria for axial SpA (in patients with back pain $\geq 3$ months and age at onset $< 45$ years)

| Sacroiliitis on imaging<br>plus<br>$\geq 1$ SpA feature                                                                                                                                                                                                                                                                                                  | or | HLA-B27<br>plus<br>$\geq 2$ other SpA features                                                                                                                                                                                                                                                                                                                              |
|----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|----|-----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| <ul style="list-style-type: none"> <li>• inflammatory back pain</li> <li>• arthritis</li> <li>• enthesitis (heel)</li> <li>• uveitis</li> <li>• dactylitis</li> <li>• psoriasis</li> <li>• Crohn's disease/ulcerative colitis</li> <li>• good response to NSAIDs</li> <li>• family history for SpA</li> <li>• HLA-B27</li> <li>• elevated CRP</li> </ul> |    | <p>Sacroiliitis on imaging</p> <ul style="list-style-type: none"> <li>• active (acute) inflammation on MRI highly suggestive of sacroiliitis associated with SpA</li> <li>OR</li> <li>• definite radiographic sacroiliitis according to mNY criteria</li> </ul> <p>sensitivity 82.9%, specificity 84.4% (overall)<br/>sensitivity 66.2%, specificity 97.3% (imaging on)</p> |



Enthesitis



Dactylitis

### Ankylosing spondylitis

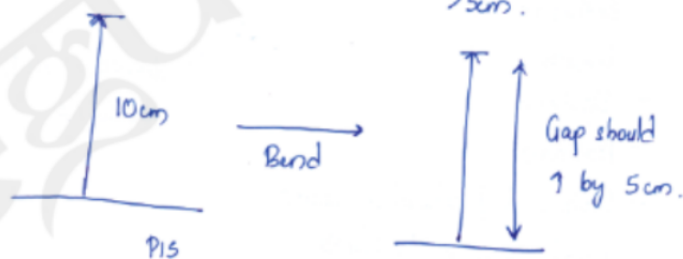
- Age < 45 years
- low backache x 3 months
- O/E - Restriction of lumbar vertebra

Shober's test → Modified Shober's test.

#### Shober's test :



#### Modified Shober's :



• HLA B<sub>27</sub> → >90% - Sm. → ↑NPV.  
Sp ↓↓.

• ↑CRP

• Lung → •ILD

• Apical fibrosis

• Extraparenchymal restriction +

• Heart → • AR +

• Dilated root of aorta

• Conduction abn.

• Neuro → • Spinal cord compression

• # → compression → Cauda equina syndrome

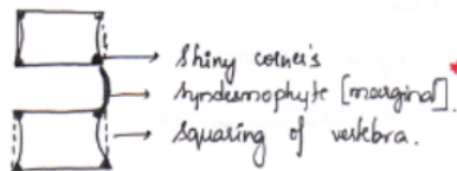
• Anterior uveitis - (MC) extraarticular manifestations  
- 33%.

• IBD +



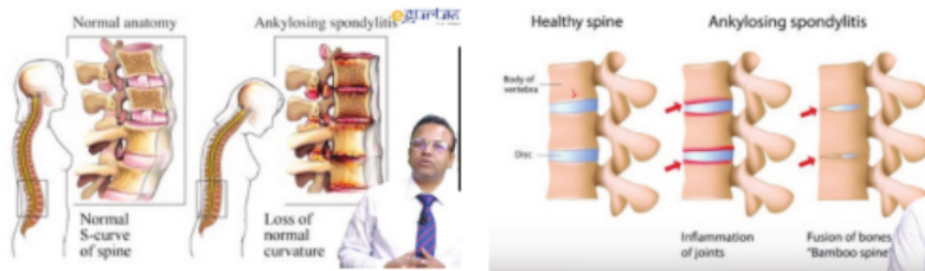
X-ray :

- B/L symmetrical<sup>\*</sup> sacroiliitis
- Bamboo spine



- Dagger sign - AP views.
  - Ossification of interspinous ligaments.
- MRI - sacroiliitis - early  
[10 yrs before Xray].
- STIR - fat suppression.

Ferguson view - Better for sacroiliac joint.

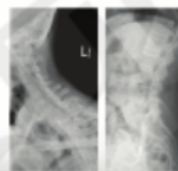


### Ankylosing Spondylitis: Definition

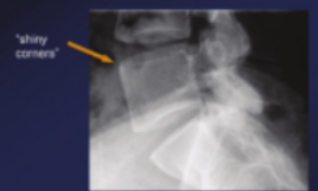
- A chronic, systemic, inflammatory disorder involving the SI joints, spine & often hips
- Axial joints are always involved, peripheral joints are frequently affected
- Characterized by "inflammatory" back pain, loss of spinal mobility
- In severe cases, extensive fusion (ankylosis) of vertebrae can increase the risk of spinal deformity, fracture, & disability

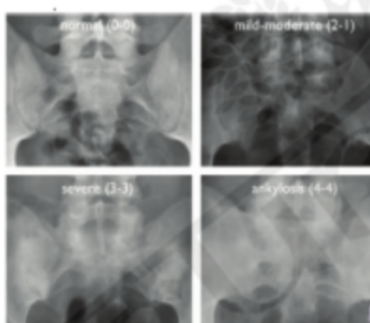
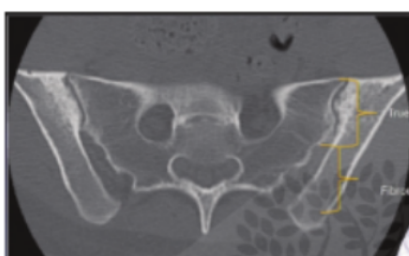
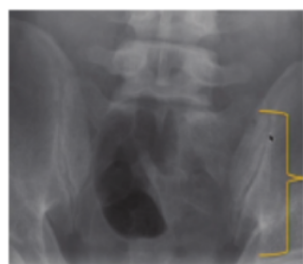


Bony fusion of vertebral bodies results in functional impairment and increases fracture risk



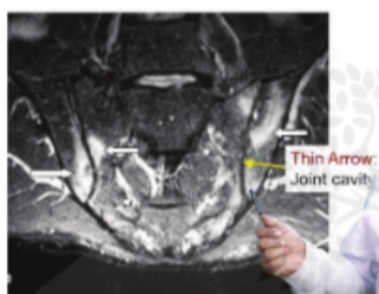
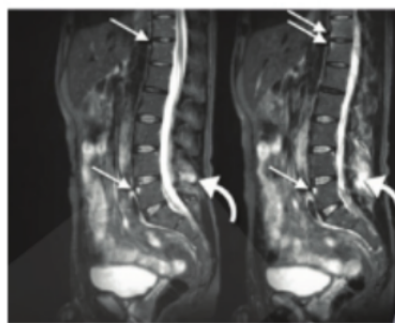
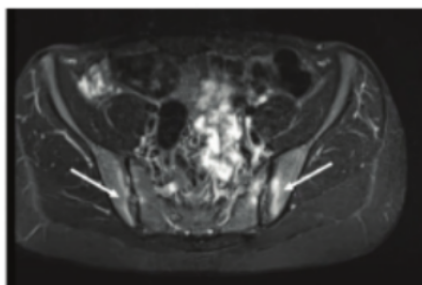
### Ankylosing Spondylitis





→ Dagger sign

STIR MRI Images



### Treatment :

- NSAIDs + Physical therapy
- Anti TNF
- IL-22/23/17 antagonists
- Mtx, Sulphasalazine, Hcq - better for peripheral ds.

X Steroids

### Exercise and PT in axial SpA/AS

- Cochrane review 2008:  
inpatient spa-exercise therapy followed by group PT > group PT > individual home-based exercises > no intervention
- practical recommendations:
  - spinal extension + deep breathing exercises twice daily
  - maintain proper posture
  - sleep on firm mattress without pillow
  - sources of information for patients: Spondyloarthritis Association of America (SAA), YouTube
- consider degree of spinal involvement:
  - avoid contact and other risk sports in patients with ankylos
  - no restrictions in non-radiographic axial SpA

### Management of active axial SpA/AS

- NSAIDs + PT are first-line therapy
- no role for conventional DMARDs
- no systemic corticosteroids, but consider local injections
- start TNFi if disease remains active on NSAIDs  
i.e. insufficient response (or intolerance) to  $\geq 2$  NSAIDs at full dose over 1 month

| BASDAI = Bath AS Disease Activity Index                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                               | ASDAS = AS Disease Activity Score                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                 |
|-----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|-----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| <ol style="list-style-type: none"> <li>1. How would you describe the overall level of fatigue/tiredness you have experienced? <b>fatigue</b></li> <li>2. How would you describe the overall level of AS neck, back or hip pain you have had? <b>back pain</b></li> <li>3. How would you describe the overall level of pain/swelling in joints other than neck, back, hips you have had? <b>peripheral arthritis</b></li> <li>4. How would you describe the overall level of discomfort you have had from any areas tender to touch or pressure? <b>enthesitis</b></li> <li>5. How would you describe the overall level of morning stiffness you have had from the time you wake up? <b>severity of morning stiffness</b></li> <li>6. How long does your morning stiffness last from the time you wake up? (0-2 hours) <b>duration of morning stiffness</b></li> </ol> | <ul style="list-style-type: none"> <li>• Parameters               <ul style="list-style-type: none"> <li>- back pain [0-10] (BASDAI Q2)</li> <li>- duration of morning stiffness [0-10] (BASDAI Q6)</li> <li>- patient global assessment [0-10]</li> <li>- peripheral joint pain/swelling [0-10] (BASDAI Q3)</li> <li>- CRP [mg/l] or ESR [mm/h]</li> </ul> </li> <li>• ASDAS-CRP<br/> <math>0.12 \times \text{back pain} + 0.06 \times \text{duration of morning stiffness} + 0.11 \times \text{patient global} + 0.07 \times \text{peripheral joint pain/swelling} + 0.58 \times \ln(\text{CRP}+1)</math><br/>               (if CRP &lt; 2 mg/l → use 2 for calculation)             </li> <li>• Current disease activity<br/>               inactive, moderate <math>\geq 1.3</math>, high <math>\geq 2.1</math>, very high <math>\geq 3.5</math> </li> <li>• Change in disease activity<br/>               clinically important <math>\geq 1.1</math><br/>               major improvement <math>\geq 2.0</math> </li> </ul> |
| 6 questions (visual analog or numerical rating scale)<br>range 0-10<br>BASDAI $\geq 4$ indicates active disease                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                       | $\text{BASDAI} = \frac{Q1 + Q2 + Q3 + Q4 + Q5 + Q6}{6}$                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                           |

IL-17 antagonists - Secukinumab → mucocut candidiasis  
 IL-12/23 antagonists - Ustekinumab

**ASAS**  
 Ankylosing Spondylitis Disease Activity Score

Back Pain (0-10)

Duration Morning Stiffness (0-10)

Patent Global (0-10)

Peripheral Pain/Tendering (0-10)

C-Reactive Protein (mg/L)

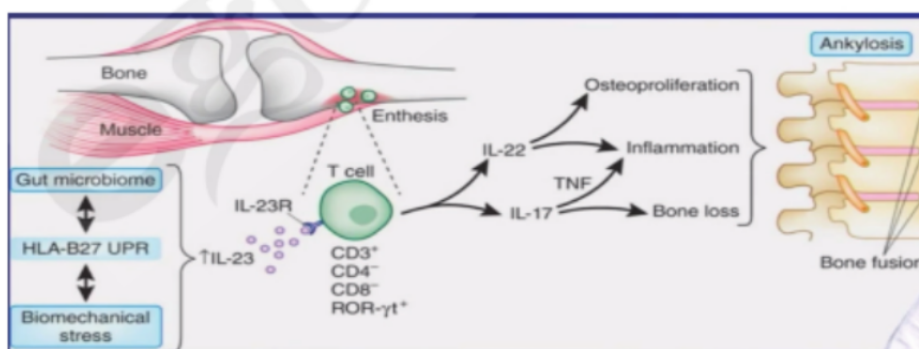
Erythrocyte Sedimentation Rate (mm/hr)

ASAS CRP

ASAS ESR



- NSAIDs + PT are first-line therapy
- no role for conventional DMARDs
- no systemic corticosteroids, but consider local injections
- start TNFi if disease remains active on NSAIDs  
 i.e. insufficient response (or intolerance) to ≥2 NSAIDs at full dose over 1 month
- monitor disease activity using BASDAI or ASDAS
- TNFi failure
  - switch to alternative TNFi OR
  - switch to IL-17A inhibitor





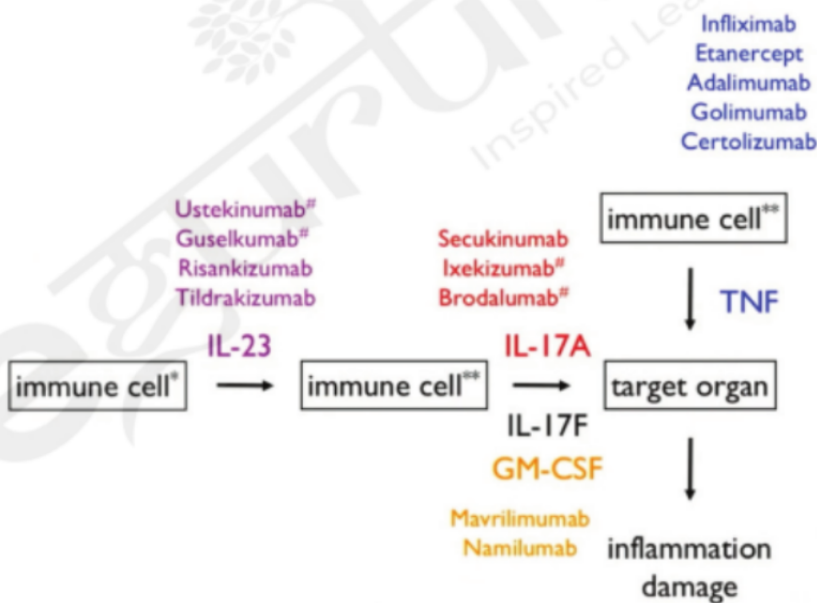
## IL17A inhibition in axial SpA/AS

### Secukinumab dosing

- approved dose for AS is 150 mg SQ monthly (± weekly loading for one month)
- anecdotal evidence that 300 mg is more effective in some patients

### Mucocutaneous candidiasis

- common SE of all IL-17A inhibitors (up to 5% in RCTs)
- typically mild-moderate and responsive to therapy  
→ treat topically and continue IL-17A antagonist



## RHEUMATOLOGY - PART-3 - ARTHRITIS

Ankylosing spondylitis  
IBD ass SpA

Reactive arthritis  
Psoriatic arthritis

Sacroiliitis - Symmetric

Asymmetric

Ligament - Anterior longitudinal  
ligament +  
Post long ligament

ALL +  
PLL ±

Symmetric vertebral  
spaces

Asymmetric  
vertebral spaces

Axial joint +++

+

Peripheral joint +

+++

Enthesitis Less

+++

Rash -

+++

Bowel inflammation +

±



## PSORIASIC ARTHRITIS

### - CASPAR criteria



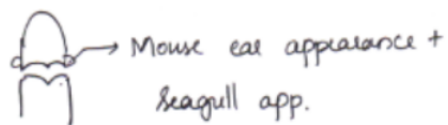
### Types :

- Oligoarticular ( $\leq 4$  joints) [ DIP, PIP. ]
- Polyarticular (like RA)
- DIP only
- Arthritis mutilans (destructive)
- Spondyloarthropathy

### CFs :

- Nail changes - Pitting  
Splitting  
Dystrophic
- DIP ⊕.

Xray :



Telescoping sign +

Asymmetric sacroiliitis.

Inv:

- CBC, LFT,  $\frac{\text{BUN}}{\text{Cr}}$

- Uric acid  $\uparrow$  [skin turnover is high]

- Rheumatoid factor  $\ominus$

$\hookrightarrow$  15% -  $\oplus$ .

- Xray - Hand & wrist Xray  
Foot & ankle "  
Lumbar/SI joint "

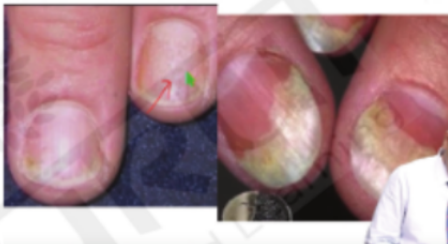
## PsA Classification: CASPAR Criteria

Inflammatory musculoskeletal disease (joint, spine, or enthesal) plus  $\geq 3$  points from the following criteria:

| Criterion                                       | Explanation                                                                                                                            | Points |
|-------------------------------------------------|----------------------------------------------------------------------------------------------------------------------------------------|--------|
| Current psoriasis                               | Current psoriasis skin or scalp disease as judged by a dermatologist or rheumatologist                                                 | 3      |
| Personal history of psoriasis                   | History of psoriasis according to the patient or a family doctor, dermatologist, or rheumatologist                                     | 1      |
| Family history of psoriasis                     | History of psoriasis in a first- or second-degree relative according to the patient                                                    | 1      |
| Psoriatic nail dystrophy                        | Typical psoriatic nail dystrophy (eg, onycholysis, pitting, hyperkeratosis) according to observation during dermatological examination | 1      |
| Negative test for rheumatoid factor             | Based on reference range or local laboratory; any testing method except latex, with performance for RF or rheumatoid factor            | 1      |
| Dactylitis                                      | Swelling of an entire digit according to observation on dermatological examination                                                     | 1      |
| History of dactylitis                           | According to a rheumatologist                                                                                                          | 1      |
| Radiologic evidence of psoriatic bone formation | On defined radiations, new joint margins (including enthesophytes, erosions) on plain radiographs of hand or foot                      | 1      |



### Clinical Presentation of Moderate to Severe Fingernail Psoriasis



### Psoriatic Arthritis





Sausage digits - entire digit is swollen → PsA > Reiter's

**PASI SCORE: 10.7**

| Head and Neck      |            |
|--------------------|------------|
| SKIN AREA INVOLVED | 2 palms    |
| AREA SCORE         | 2          |
| REDNESS            | 3          |
| THICKNESS          | 4          |
| SCALE              | 1          |
| <b>TOTAL:</b>      | <b>1.6</b> |

| Trunk              |            |
|--------------------|------------|
| SKIN AREA INVOLVED | 2 palms    |
| AREA SCORE         | 1          |
| REDNESS            | 3          |
| THICKNESS          | 0          |
| SCALE              | 2          |
| <b>TOTAL:</b>      | <b>1.5</b> |



- "Pencil-in-cup" deformity
- Resorption of terminal phalanges





- Skin + Arthritis → Methotrexate.
- Apremilast → PDE-4 inhibitor
- IpA → Anti-TNF
- IL-17 antagonist - Secukinumab
- IL-12/23 " - Ustekinumab

- Skin + Asthma → Methotrexate
- Apremilast → PDE-4 inhibitor
- SpA → Anti-TNF
- IL-17 antagonist - Secukinumab
- IL-12/23 " - Ustekinumab

## Reiter's (Reactive) arthritis

Triggers: <sup>M.T.</sup> Chlamydia  
<sup>M.C. G. cause</sup> Shigella  
 Salmonella  
 Campylobacter

Yersinia  
 Ureaplasma urealyticum  
 Mycoplasma genitalium  
 HIV.

X Gonococci

Triad:



Skin: - Palms & Soles - Keratoderma blennorrhagica  
 - Penile - Balanitis circinata

Dr:

- Urethritis - Doxycycline.
- NSAIDs
- Anti-TNF
- IL-17, IL-12/23 antagonists.

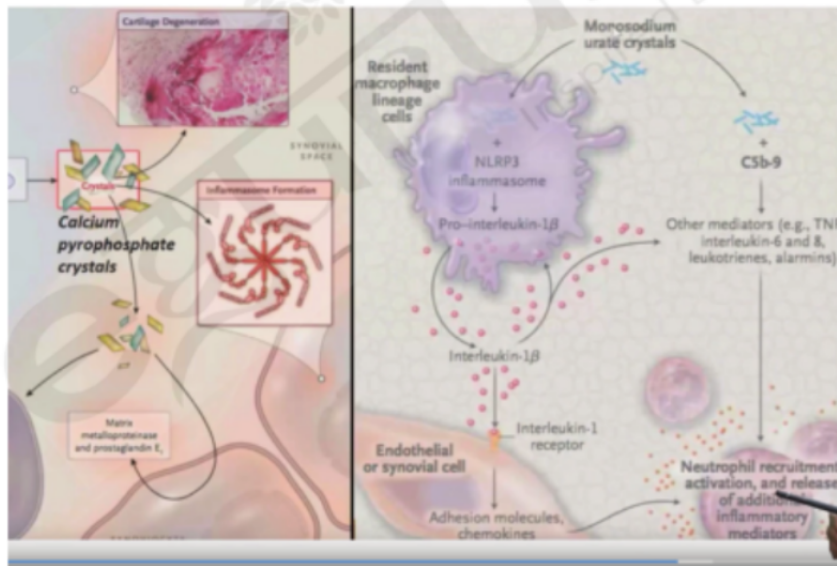
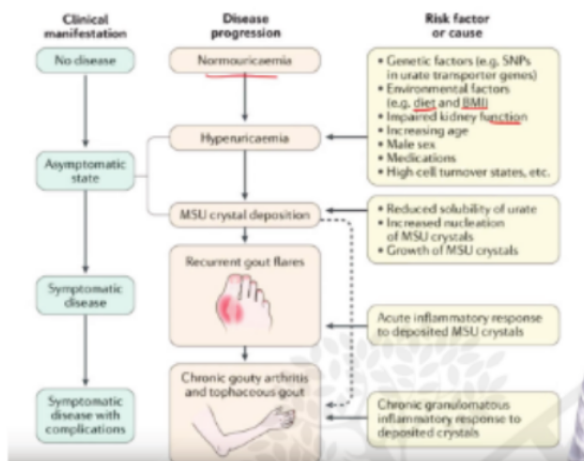
### ReA Mucocutaneous Manifestations

- Circinate balanitis
- Conjunctivitis
- Keratoderma blennorrhagica
- Painless oral ulcers



## Gout

### Pathophysio :





## Gout

- Precipitation of monosodium urate crystals in joints.
- Neutrophils - engulf crystals → enzymes  
↓  
Pain → inflammation

- ↓ Temp.

- Monoarticular / oligoarticular ( $\leq 4$  joints)

- 1st joint - 1st metatarsophalangeal joint  
↓  
Podagra.

- Night → severe attack of pain.

- Trigger → Alcohol / Stress / Drugs / Surgery.

O/E - Joint → Swollen  
Erythema (+)  
Tender  
Tophi.

Tophi → deposition of monosodium urate crystals  
↓  
synovium, subchondral bone, olecranon,  
extensor surfaces

- Serum uric acid
  - ↳ during acute flare → ↓
  - ↳ after flare → high.

- Synovial fluid analysis:

↳ TLC → ↑.

- Crystals - Na urate  
Negative birefringence  
Needle shaped.

Under polarised light → Parallel light → Yellow. → Negative birefringent.

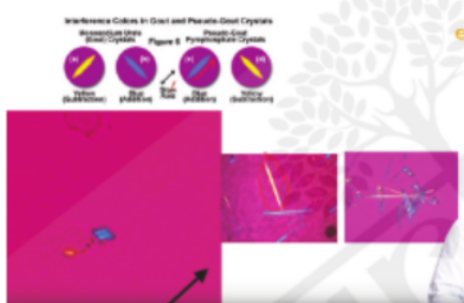
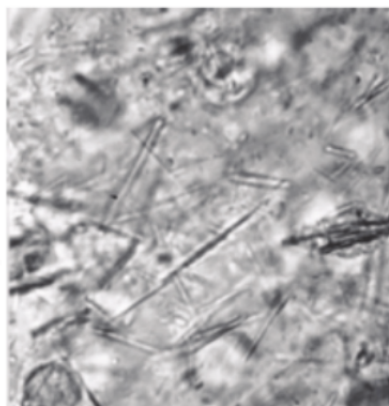
Gout - uric acid ↑

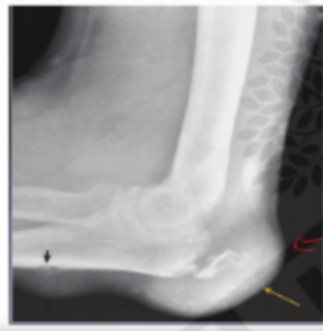
Over production

Under excretion

• Urine uric acid ↑  
>800 mg/day

mc  
↓  
<800 mg/day.





### Dual Energy CT – History & Use

Noninvasive differentiation of urinary stone compositions in patients with nephrolithiasis using dual-source dual-energy CT

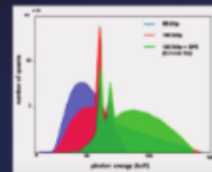
**Description:** Dual-energy CT using a full-spectrum CT scanner involves differentiating of urinary stones with greater than 95 percent accuracy. The second-generation dual-source CT scanner includes an additional filter in the high-energy x-ray tube (or filter) to increase the kVp spectra separation. This, in turn, increases the difference in dual-energy CT number ratio between stone types. Differentiation between four groups of stone types may be achieved using the second-generation dual-source CT scanner.

- DECT conceived in 1975.
- Technology recently applied to the evaluation, diagnosis, and management of gout in the musculoskeletal system.



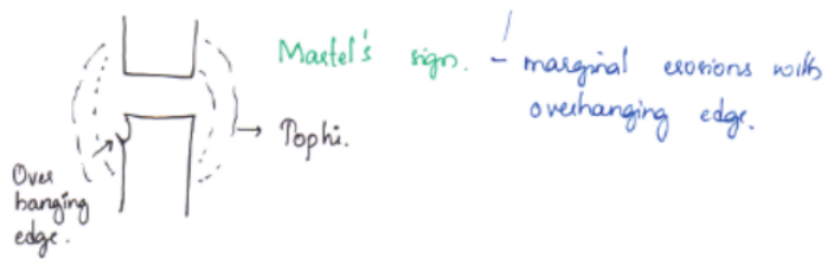
### DECT: Technique

- DECT uses two x-ray tubes with different peak kilovoltages: **80 and 140 kVp**
- **No Contrast**



These are used to acquire two CT image sets of a specific anatomic region **simultaneously**. This eliminates misregistration or errors that can occur due to movement if the imaging is sequentially.

The chemical composition of the scanned material can be differentiated.



- Dual energy CT → 80 & 140 kVP.  
     ↳ Differentiation of stones  
     GREEN → GOUT

## RHEUMATOLOGY - PART-4 ARTHRITIS

Re:

Acute attack:

- NSAIDs - DOC
- Steroids → intra articular inj/oral
- Colchicine → ↓ neutrophil chemotaxis  
↓ release of cytokine  
↓  
↓ Inflammation

High dose: e/e - Diarrhea  
↳ sometimes - bloody diarrhea

- Rest, cold compress

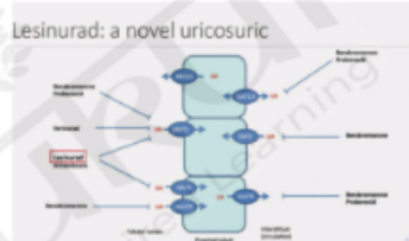
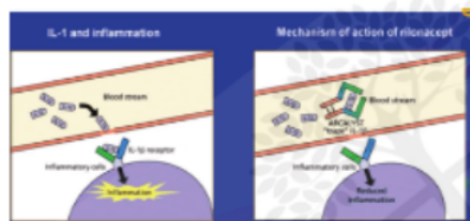
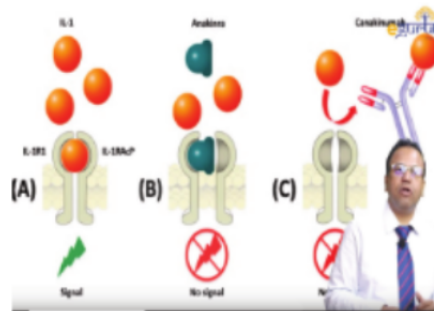
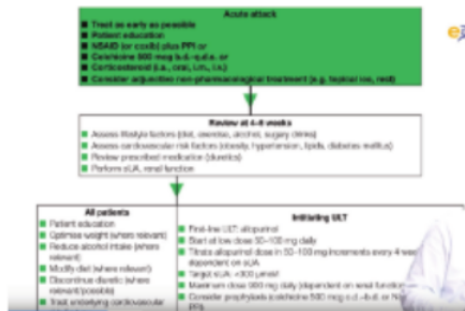
Urate lowering therapy - after acute attack subsides

DOC Allopurinol

Febuxostat

Probenecid

Pegloticase [Urican] → ↑ uric acid solubility.



**ORIGINAL ARTICLE**

**Cardiovascular Safety of Febuxostat or Allopurinol in Patients with Gout**

William B. White, M.D., Kenneth G. Saag, M.D., Michael A. Becker, M.D., Jeffrey S. Borer, M.D., Philip B. Gornick, M.D., Andrew Whellan, M.D., Barbara Frant, M.S., Maje Goffin, M.D., and Jennifer Gombart, M.D., Ph.D., for the CARET Investigators\*

*\* See text for definitions of abbreviations and acronyms.*

**Table 2. Major Safety End Points (Pooled Intention-to-Treat Analysis)\***

| End Point                                                                                                                                                   | Febuxostat (N=1206) | Allopurinol (N=1092) | Hazard Ratio (95% CI) | P Value |
|-------------------------------------------------------------------------------------------------------------------------------------------------------------|---------------------|----------------------|-----------------------|---------|
| Primary end point: composite of cardiovascular death, nonfatal myocardial infarction, nonfatal stroke, urgent medical attention for cardiovascular symptoms | 115 (9.5)           | 101 (9.4)            | 1.01 (0.87-1.17)      | 0.92    |
| Secondary end points                                                                                                                                        |                     |                      |                       |         |
| Cardiovascular death                                                                                                                                        | 119 (9.8)           | 106 (9.7)            | 1.04 (0.89-1.21)      | 0.58    |
| Nonfatal myocardial infarction                                                                                                                              | 111 (9.1)           | 104 (9.5)            | 0.93 (0.79-1.10)      | 0.43    |
| Nonfatal stroke                                                                                                                                             | 71 (5.8)            | 70 (6.4)             | 1.06 (0.79-1.43)      | 0.68    |
| Urgent medical attention for cardiovascular symptoms                                                                                                        | 119 (9.8)           | 106 (9.7)            | 1.04 (0.89-1.21)      | 0.48    |
| Composite of cardiovascular death, nonfatal myocardial infarction, or nonfatal stroke                                                                       | 119 (9.8)           | 106 (9.7)            | 1.04 (0.89-1.21)      | 0.53    |
| Death from any cause                                                                                                                                        | 141 (11.6)          | 128 (11.7)           | 1.02 (0.89-1.17)      | 0.78    |

**Febuxostat/Uloric 'Black Box' Warning**

- Studies show febuxostat has an increased risk of death compared with allopurinol
- Febuxostat is now approved for use only in people who try allopurinol first
- Pay attention to symptoms of heart attack and stroke while taking febuxostat

NSAIDs / low dose colchicine

can be used along with urate lowering therapy  
to prevent / treat acute flare.

Target: Uric acid < 6mg/dL

Tophi: " < 5mg/dL.

New medication in Acute attack:

Anakinra - IL-1 receptor blocker.

Canakinumab (best)  
Rilonacept } IL-1 blocker.

Lesinurad → acts on URAT-1  
novel uricosuric.

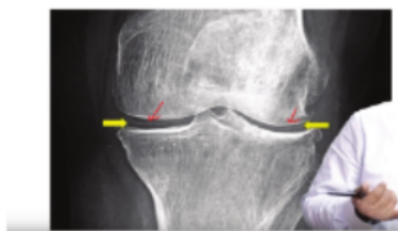
s/e : • Atlopurinol - HLA-B5801  
↓  
skin rash

• Febuxostat - ↑ CVS risk.



### Pseudo - gout :

- Deposition of Calcium pyrophosphate dehydrogenase.
- MC joint - Knee
- Oligoarticular.
- Associated with :
  - Hyperparathyroid
  - Familial hypocalcaemic hypercalcaemia
  - Haemochromatosis\*
  - Hypothyroidism.
  - ↓ Mg, ↓  $PO_4$
  - Trauma
  - Rheumatoid arthritis
  - Use of bisphosphonates\*
- Synovial fluid analysis:
  - TLC ↑↑ (>2000/cf)
  - Positive birefringent crystals [parallel light → Blue]
  - Rhomboid shape.
- Xray: Calcification ⊕.



R:

Acute inflammation:

- NSAIDs
- Colchicine
- Steroids.

Treat underlying cause.

DISH → Diffuse idiopathic skeletal hyperostosis

- Ankylosing hyperostosis
- >50 yrs
- Thoracic, lumbar, cervical
- Association - **DM**.
- D/D - Ankylosing spondylitis
- Anterior longitudinal ligament involved } Restricted  
Osteophytes } vertebra  
↓ +  
Spinal stenosis. } Pains.

Ankylosing spondylitis

Age: &lt; 40yrs

Sacroiliitis - B/L  
symmetrical

↑ ESR, ↑ CRP

⊖

Bony ankylosis ⊕

Disc space - Narrow

Ankylosing hyperostosis (DISH)

&gt; 50yrs

Spared.

⊖

Osteophytes ⊕

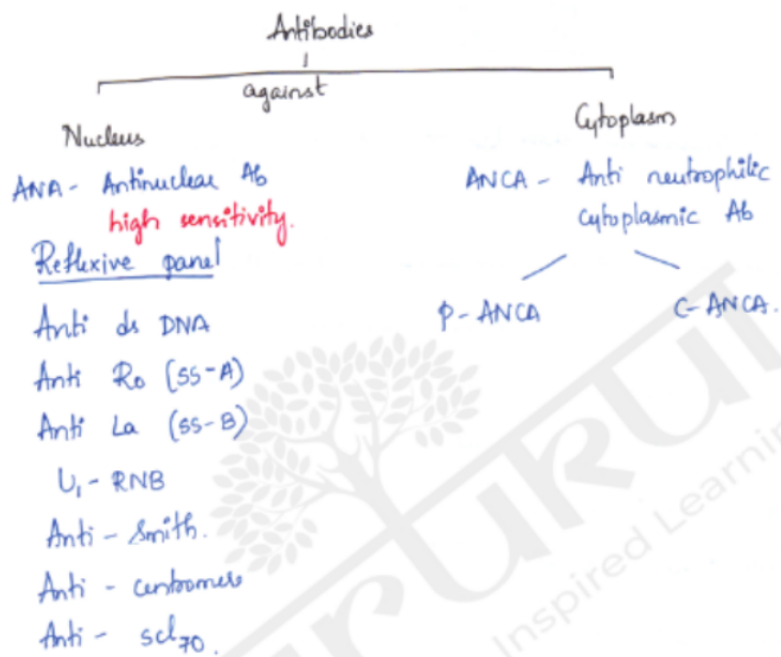
⊕

[4 consecutive vertebrae]  
fused.  
maintained.

Xray: Flowing wax sign.



## TESTS IN RHEUMATOLOGY - ANTIBODIES



### ANA ⊕

- |                               |                                    |
|-------------------------------|------------------------------------|
| - SLE → <i>&gt;95% → ↑NPV</i> | - Rheumatoid arthritis             |
| - Scleroderma                 | - Vasculitis                       |
| - Sjogren's                   | - Subacute bacterial endocarditis. |
| - Myositis                    |                                    |
| - Chronic active hepatitis    |                                    |
| - Infectious mononucleosis    |                                    |
| - Leprosy                     |                                    |

ANA ⊕ → • ELISA

• Immuno-fluorescent assay. - (better)

> 1:80 → Significant.

\* Rheumatoid factor ⊕ → RA & Sjogren's.

ANA - SLE, Scleroderma, Sjogren's.

Anti-centromere - CREST syndrome

Anti-scl-70 - Diffuse systemic sclerosis

U<sub>1</sub>-RNP - Mixed connective tissue disease

Anti-Ro (SS-A) } Sjogren's, SLE.

Anti-La (SS-B) }

LKM antibodies

LKM-1 - Autoimmune hepatitis, HCV

LKM-2 - Drug induced hepatitis

LKM-3 - HDV

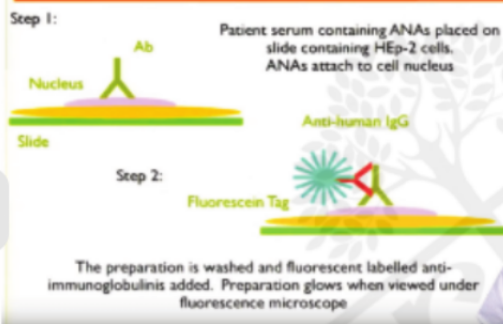
Antimitochondrial - Primary biliary cholangiopathy.

Anti-thyroglobulin - Post partum thyroiditis  
Hashimoto's thyroiditis

Anti-mitochondrial Abs - Hashimoto's thyroiditis  
Post partum thyroiditis

- ASCA Ab - Crohn's ds  
(Anti *saccharomyces cerevisiae* Ab)
- Anti-smooth muscle Ab - Autoimmune hepatitis
- Anti-Jo<sub>1</sub> - Myositis
- p-ANCA - Small vessel vasculitis

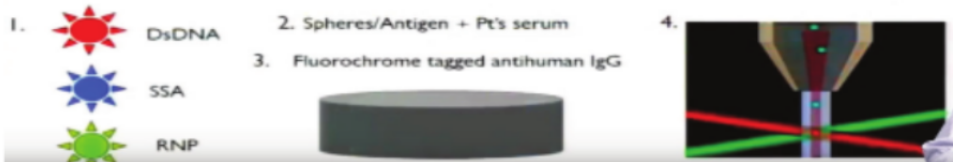
#### FANA Technique



- ↳ Granulomatosis with polyangiitis [GPA]
- ↳ Microscopic polyangiitis [MPA]
- ↳ Eosinophilic granulomatosis with polyangiitis [EGPA]
- ↳ Ulcerative colitis

#### Multiplexed Method for Detecting Autoantibodies

1. Autoantigen is fused to colored microspheres
2. Pt serum is then incubated with microspheres; if autoantibodies are present these will attach to the autoantigen/sphere complex
3. Antihuman IgG/fluorochrome tag then added
4. Microsphere then placed in flow cytometry column that uses two lasers, one to detect color of sphere and one to identify the presence of the antibody/fluorochrome complex
5. Results reported as a number not titer



## SLE

ANA - >95% - sm  
5% - sp.

### Immunofluorescence :

Peripheral (Rim) - anti ds-DNA  
Speckled - Scleroderma, mixed connective  
tissue ds (MCTD), Sjogrens  
Nucleolar - Scleroderma  
Centromere - CREST

### AutoAB

#### Seen in

Anti- dsDNA - 40% SLE  
Very specific  
Nephritis\*  
Anti- Smith - 25% SLE  
Extremely specific  
Anti- RNP - MCTD, SLE  
Anti- Ro/La - Cutaneous SLE,  
Neonatal SLE  
Sjogren's

Anti dsDNA :

- Correlates with disease severity
- Nephritis\*

Anti Smith :

- Most specific\* [Most bn → ANA]
- 25% - bn.

U<sub>1</sub>-RNP :

- Mixed connective tissue disease (MCTD)

Overlap syndrome :

> 1 connective tissue disease, each fulfilling the classification criteria.

- SLE, Scleroderma, Myositis, RA

Overlap syndrome.

↓

Overlap syndrome + U<sub>1</sub>RNP (↑) → MCTD.

Anti-histone : Drug induced lupus.

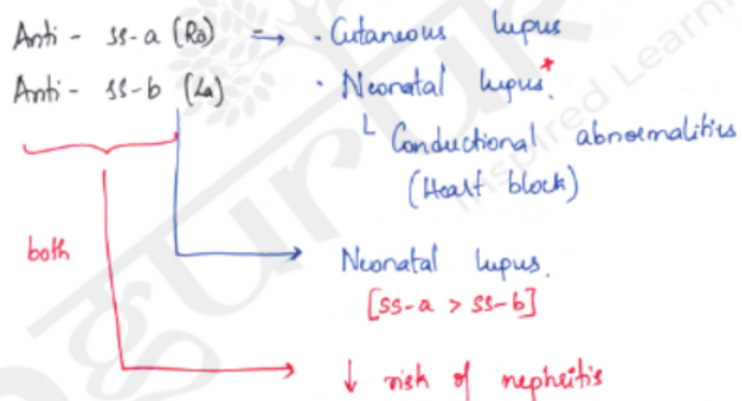
- Hydralazine
- Sulphonamide
- Procainamide
- HC-TZD.
- Anti-TNF



### Drug induced lupus :

- Transient - on stopping the drug  
↓  
anti-histone titres ↓
- HLA-DR<sub>4</sub>\*
- Anti-dsDNA - negative.

Anti-Ribosomal-P : CNS lupus\*  
Hepatitis ⊕

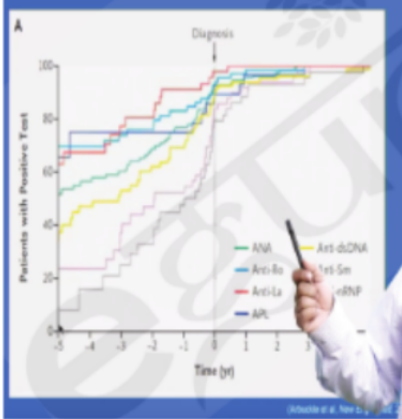


Antiphospholipid Abs -

- ↑ Thrombosis
- fetal loss

| ANTIBODY                                          | PREVALENCE, % | ANTIGEN RECOGNIZED                                                            | CLINICAL UTILITY                                                                                                                                                                                                        |
|---------------------------------------------------|---------------|-------------------------------------------------------------------------------|-------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| Antinuclear antibodies                            | 98            | Multiple nuclear                                                              | Best screening test; repeated negative tests by immunofluorescence SLE unlikely                                                                                                                                         |
| Anti-dsDNA                                        | 70            | DNA (double-stranded)                                                         | High titers are SLE-specific and in some patients coincide with disease activity, nephritis, vasculitis. Onslide immunofluorescence is more specific for SLE than ELISA methods.                                        |
| Anti-Sm                                           | 25            | Protein complexed to 6 species of nuclear U1 RNA                              | Specific for SLE; no definite clinical correlations; most patients also have anti-RNP; more common in blacks and Asians than whites                                                                                     |
| Anti-RNP                                          | 40            | Protein complexed to U1 RNA                                                   | Not specific for SLE; high titers associated with syndromes that have overlap features of several rheumatic syndromes including SLE; more common in blacks than whites; correlates with high IFN-induced gene signature |
| Anti-Ro (SSA)                                     | 30            | Protein complexed to H1 RNA, primarily 60 kDa and 52 kDa                      | Not specific for SLE; associated with sicca syndrome, predisposes to subcutaneous lupus, and to neonatal lupus with congenital heart block; associated with decreased risk for nephritis                                |
| Anti-La (SSB)                                     | 10            | 47 kDa protein complexed to H1 RNA                                            | Usually associated with anti-Ro; associated with decreased risk for nephritis                                                                                                                                           |
| Anti-histone                                      | 70            | Histones associated with DNA (in nucleosome, chromatin)                       | More frequent in drug-induced lupus than in SLE                                                                                                                                                                         |
| Antiphospholipid                                  | 50            | Phospholipids, $\beta_2$ glycoprotein 1 ( $\beta_2$ G1) cofactor, prothrombin | Three tests available—ELISAs for cardiolipin and $\beta_2$ G1, sensitive prothrombin time (DVT); for lupus anticoagulant; predisposes to clotting, fetal loss, thrombocytopenia                                         |
| Anterythrocyte                                    | 60            | Erythrocyte membrane                                                          | Measured as direct Coombs test; a small proportion develops overt hemolytic                                                                                                                                             |
| Antiplatelet                                      | 30            | Surface and altered cytoplasmic antigens on platelets                         | Associated with thrombocytopenia, but sensitivity and specificity poor; this is not a useful clinical test                                                                                                              |
| Antineuronal (includes anti-glutamate receptor 2) | 60            | Neuronal and lymphocyte surface antigens                                      | In some series, a positive test in CSF correlates with auto                                                                                                                                                             |
| Antiribosomal P                                   | 20            | Protein in ribosomes                                                          | In some series, a positive test in serum correlates with psychosis due to CNS focus                                                                                                                                     |

### Autoantibodies prior to SLE



### Extractable Nuclear Antibodies

| ENA antibody         | Abbreviation              | Disease Association(s)            |
|----------------------|---------------------------|-----------------------------------|
| Sjogren's syndrome A | SSA (Ro)                  | SLE, Sjogren's                    |
| Sjogren's syndrome B | SSB (La)                  | Sjogren's, SLE                    |
| Double stranded DNA  | DsDNA                     | SLE                               |
| Ribonuclear protein  | RNP                       | SLE MCTD (high titer in relation) |
| Smith                | Sm (not SM smooth muscle) | SLE                               |
| Scleroderma 70       | SCL 70                    | Scleroderma                       |
| Chromatin            |                           | SLE                               |
| Histone              |                           | SLE                               |

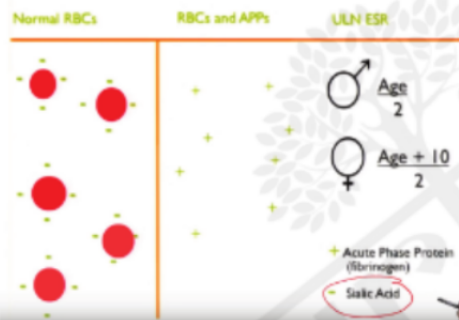
### Erythrocyte Sedimentation Rate: Methodology

#### Westergren method:

- Whole blood anticoagulated with  $K_3EDTA$
- Placed in 200 mm long, 2.5 mm wide column
- Measure distance RBC column falls in 1 hr
- Recorded in mm/hr



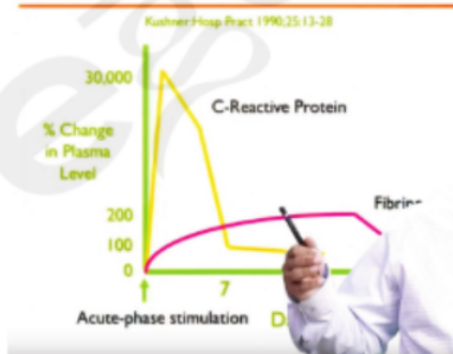
### Erythrocyte Sedimentation Rate



### Erythrocyte Sedimentation Rate

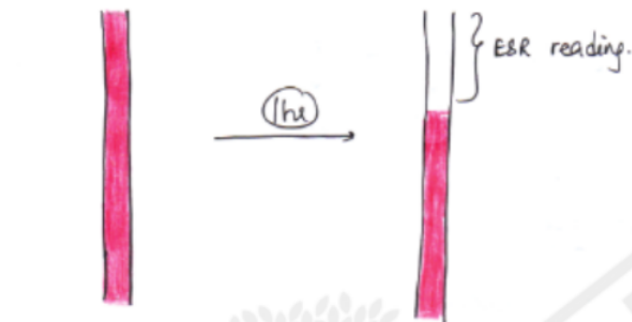


### Time Course of Acute Phase Reactants



ESR

Westergren's method :



sulfic acid  
  
 due to negative charge  $\rightarrow$  repel each other & do not stick.

$$\textcircled{N} \text{ ESR} \rightarrow \begin{matrix} \text{♂} \rightarrow \frac{\text{Age}}{2} \\ \text{♀} \rightarrow \frac{\text{Age} + 10}{2} \end{matrix}$$

Acute phase proteins (fibrinogen)  $\rightarrow$  attach to RBC  $\Rightarrow$  sink the RBC faster  
 $\downarrow$   
 $\therefore \uparrow \text{ESR}$

In SLE : Gamma globulin  $\uparrow$   
 $+++$

$\therefore \text{SLE} \rightarrow \uparrow \text{ESR}$

In SLE:

If no inflammation  $\rightarrow$  CRP may be  $\odot$ .

$\therefore$  Discrepancy b/w ESR & CRP  
 $\downarrow$                        $\downarrow$   
 $\uparrow$                        $\odot$

Important HLA:

- HLA B<sub>5</sub>  $\rightarrow$  Behcet's  
Ulcerative colitis  
Polycystic kidneys
- HLA DR<sub>1</sub> / DR<sub>4</sub>  $\rightarrow$  Rheumatoid arthritis
- HLA DR<sub>3</sub> / DR<sub>4</sub>  $\rightarrow$  Type-1 DM
- HLA - DR<sub>2</sub>  $\rightarrow$  Narcolepsy  
Multiple sclerosis  
Good Pasture's ds
- HLA - DR<sub>5</sub>  $\rightarrow$  Pernicious anemia.  
Hashimoto's thyroiditis  
TIA
- HLA - DR<sub>7</sub>  $\rightarrow$  Congenital adrenal  
hyperplasia

HLA - A<sub>29</sub> → Birdshot chorioretinitis

HLA - DQ<sub>2</sub> → Celiac's ds

HLA - DR<sub>3</sub> → SLE, Sjogren's, Celiac ds / Dermatitis  
Grave's ds, herpesiformis  
Type I DM

HLA - DR<sub>4</sub> → Drug induced lupus.



## VASCULITIS - PART 1

### Chapel Hill classification:

Large vessel : . Temporal arteritis [Giant cell arteritis]  
 . Takayasu " } Granulomas present.

Medium : . Polyarteritis nodosa (PAN)  
 . Kawasaki's vasculitis → Type II hypersensitivity

Small :

#### ANCA associated

Microscopic polyangiitis (MPA)

Wegener's (Granulomatosis with polyangiitis) (GPA) } Granulomas ⊕.

Churg Strauss (Eosinophilia with granulomatosis with polyangiitis) }  
 [EGPA]

#### Immune complex type

Henoch - Schonlein purpura (IgA vasculitis)

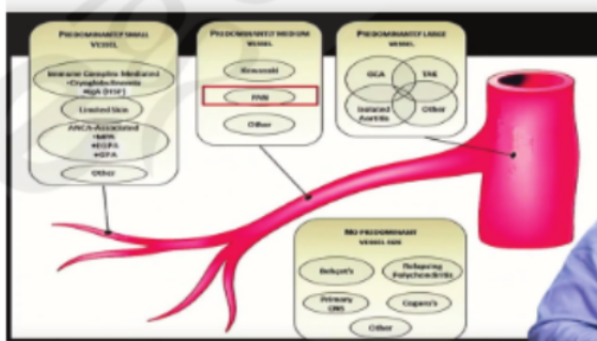
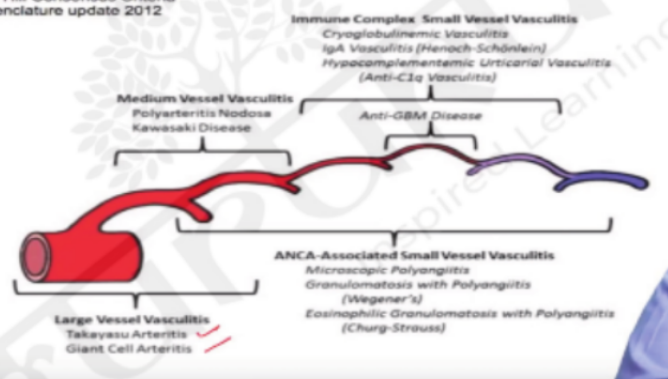
Cryoglobulinemia.

No predominant vessel size : . Behcet's  
 . Primary CNS  
 . Relapsing polychondritis  
 . Cogan's

| A. Large vessels                                                                  | B. Medium vessels                                                                 | C. Small vessels                                                                  |
|-----------------------------------------------------------------------------------|-----------------------------------------------------------------------------------|-----------------------------------------------------------------------------------|
| Aorta and its major branches                                                      | Main visceral arteries and their initial branches                                 | Intraparenchymal arteries, arterioles, capillaries                                |
|  |  |  |

## Classification of Vasculitis

Chapel Hill Consensus Criteria  
Nomenclature update 2012





### Temporal arteritis

- Giant cell arteritis
- Age  $> 50$  yrs\* [never below 50]
- F:M = 3:1
- (m.c) vessel  $\rightarrow$  superficial temporal A.
- CFs:

Headache  $\rightarrow$  (m.c) - New onset headache  
Scalp tenderness (+)

Visual abn - Post ciliary A / Ophthalmic A inv  
 $\hookrightarrow$  Anterior ischemic optic neuropathy (AION)  
 $\hookrightarrow$  irreversible  
 \* Altitudinal field defect

Jaw claudication

Constitutional symptoms - fever, myalgia, fatigue.

Polymyalgia rheumatica\* - 50% - proximal myalgia  
neck stiffness

ESR, CRP  $\Rightarrow$  ( $\uparrow\uparrow$ )  
 $\hookrightarrow$  m - 96%.

### Temporal arteritis

- Giant cell arteritis
- Age  $> 50$  yrs\* [never below 50]
- F:M = 3:1
- (m.c) vessel  $\rightarrow$  superficial temporal A.
- CFs:

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 $\hookrightarrow$  Anterior ischemic optic neuropathy (AION)  
 $\hookrightarrow$  irreversible  
 \* Altitudinal field defect

Jaw claudication

Constitutional symptoms - fever, myalgia, fatigue.

Polymyalgia rheumatica\* - 50% - proximal myalgia  
neck stiffness

ESR, CRP  $\Rightarrow$  ( $\uparrow\uparrow$ )  
 $\hookrightarrow$  m - 96%.

### Patho:

Disease starts → transmural → Occludes lumen  
from tunica adventitia (panvasculitis)

↳ Vessels without / small adventitia → not involved.

↓  
Intradural vessels not involved.

Temporal arteritis doesn't involve - Renal A.  
Pulmonary A.

- Temporal A biopsy → >1.5 cm is removed.

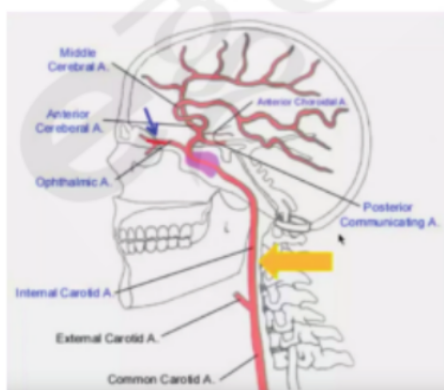
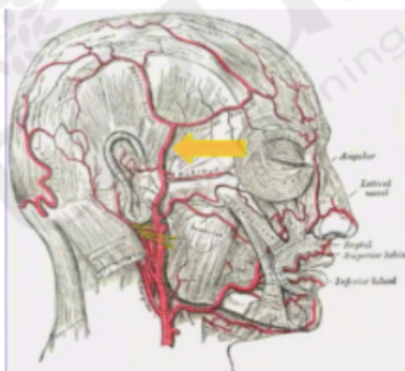
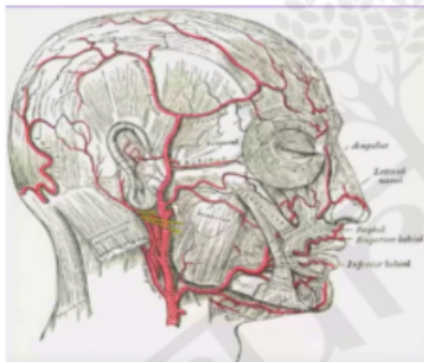


TABLE 1

#### Symptoms in giant cell arteritis\*

| Category                                                                   | Symptoms                                                                                                                                                       |
|----------------------------------------------------------------------------|----------------------------------------------------------------------------------------------------------------------------------------------------------------|
| Symptoms due to involvement of cranial vessels                             | Headache<br>Jaw claudication (pain on chewing)<br>Scalp tenderness<br>Loss of vision<br>Abnormalities of the temporal artery (pain, nodules, absence of pulse) |
| Symptoms due to involvement of great vessels (aorta and branches of aorta) | Claudication of extremities (especially arm)                                                                                                                   |
| Symptoms due to systemic inflammation                                      | Fever, night sweats, weight loss                                                                                                                               |
| Polymyalgia rheumatica                                                     | Mainly proximal myalgia and stiffness of the neck and shoulder and pelvic girdles                                                                              |

### Temporal arteritis (Giant Cell arteritis)

Female > male

Fifty and older

Fifty ESR > 50 mm/h

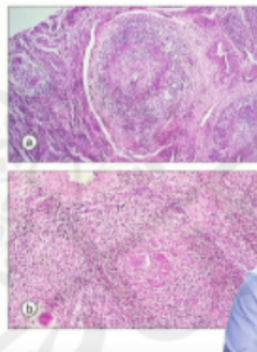
Four HLA-DR4

Firm temporal artery

Face => Pain (jaw claudication) and visual loss

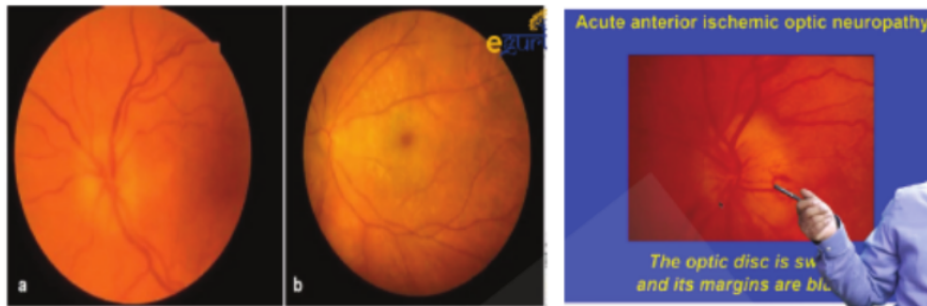
Flu-like illness (associated with Polymyalgia rheumatica)

Fever



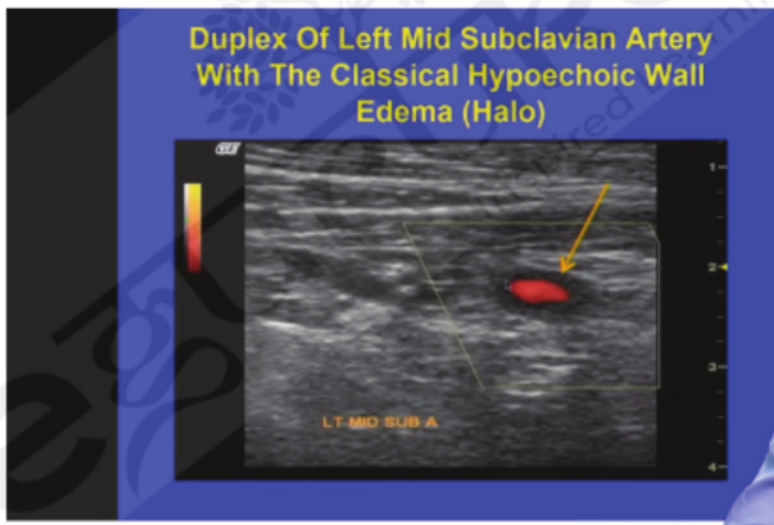
- Visual abn
- ANON
- Diplopia
- Central retinal A occlusion
- X CRVO
- Amaurosis fugax

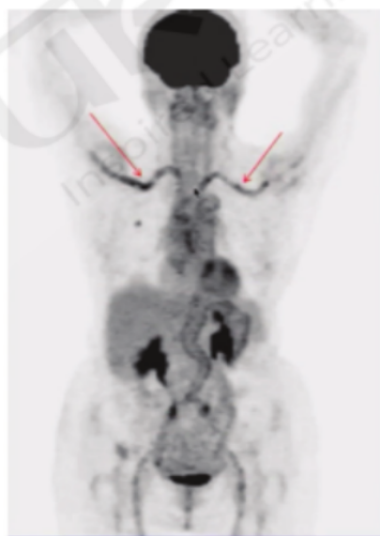
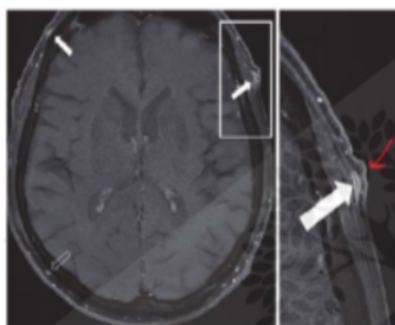
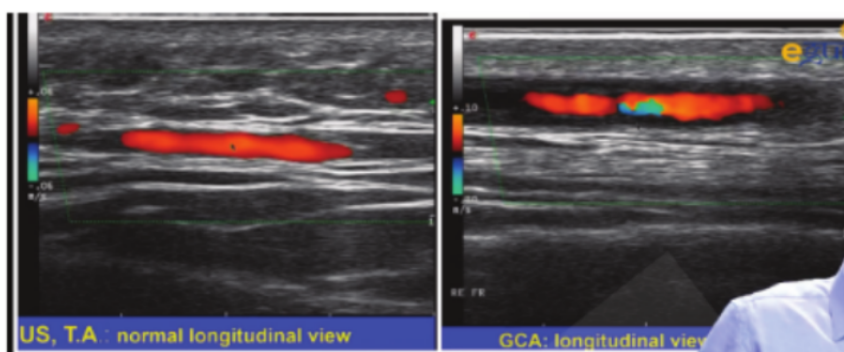
As it is irreversible (ANON) → even on suspicion of temporal arteritis, steroids should be started.



Dev:

- Gold std - Temporal A biopsy
- USG - Doughnut sign / Halo sign





### Classification criteria

- Age > 50 yrs
- New-onset headache
- Temporal A abnormalities
- ESR > 50 mm/hour
- Histologic evidence of arteritis on temporal A biopsy.

### Polymyalgia rheumatica

50f → previous temporal arteritis

5-15f → develop " "

CK, muscle biopsy → (N)

Joint & soft tissue - problem.

- Criteria:
- Morning stiffness > 45 min
  - Rheumatic factor / anti CCP Abs negative
  - Pelvic girdle pain / ↓ hip mobility
  - No other painful joint
  - USG: inflammatory changes in both shoulders
  - USG: inflammatory changes in at least one shoulder & hip joint.

Age > 50 yrs

Pain & stiffness → maximum in mornings

Neck, shoulder, Hip, thigh pain

CRP ↑↑, ESR ↑± [as fibrinogen rise takes time]

NSAIDs - poor response

Ck - ⊕.

Δ<sup>ns</sup>: usg

Re: Steroids - dramatic response.

Do of temporal arteritis:

- Steroids - should be given before investigation as AION → irreversible.
- $\downarrow$  60mg/day - high dose. → Taper every 2 weeks by 10mg.  
Prednisolone.
- Biopsy - Positive → Diagnosis  
Negative → opposite temporal A biopsy.  
- usg / MRI
- Tocilizumab. - IL-6 antagonist.



### Sequels of Temporal arteritis :

- Aortitis → Dissection of aorta
- Thoracic aneurysms

[On tapering steroids → widening is seen]

### TAKAYASU

- Non specific aortitis / Pulseless disease / Aortic arch syndrome

Age < 40 yrs

F:M = 8:1

(MC) vessel → **subclavian . A.**

CFs: fever, fatigue, wt loss, arthralgia.

- Arm claudication, Raynaud's - **subclavian . A.**

O/E - Asymmetric pulses

Difference of BP > 10 mmHg b/w 2 upper limbs.

- Visual sym, syncope - **Carotid . A**

- HTN, ↑Cr - **Renal . A**

- Dizziness - **Vertebral . A.**

- Bowel ischemia - Abd pain, nausea, vomiting, bloody diarrhea - **Celiac . A / SMA/ IMA**

- AR, CCF

Criteria:

Age < 40 yrs

Claudication of extremities

↓ brachial artery pulse

BP difference > 10 mmHg

Bruit over subclavian arteries or aorta

Arteriogram abnormality

Angiogram classification:

Type I - Branches of aortic arch

IIa - Ascending aorta, aortic arch & its branches

IIb - Type IIa + descending thoracic aorta

III - Thoracic descending aorta, abdominal aorta, renal arteries or a combination

IV - Abdominal aorta, renal arteries, or both

V - Entire aorta

Japanese classification:

I - Aortic arch

II - Asc aorta

III - Desc aorta

IV - Pulm artery ← given by India

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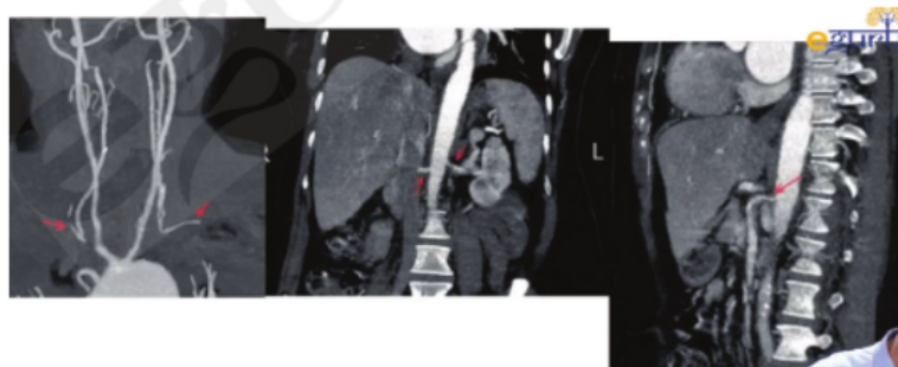
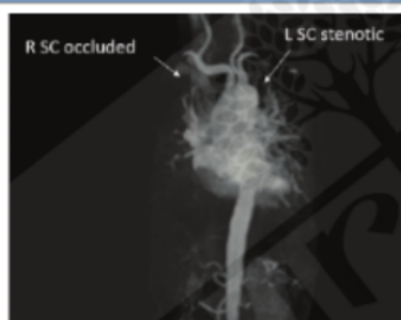
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**TABLE 356-7 Frequency of Arteriographic Abnormalities and Potential Clinical Manifestations of Arterial Involvement in Takayasu Arteritis**

| ARTERY               | PERCENTAGE OF ARTERIOGRAPHIC ABNORMALITIES | POTENTIAL CLINICAL MANIFESTATIONS                           |
|----------------------|--------------------------------------------|-------------------------------------------------------------|
| Subclavian           | 93                                         | Arm claudication, Raynaud's phenomenon                      |
| Common carotid       | 58                                         | Visual changes, syncope, transient ischemic attacks, stroke |
| Abdominal aorta*     | 47                                         | Abdominal pain, nausea, vomiting                            |
| Renal                | 38                                         | Hypertension, renal failure                                 |
| Aortic arch or root  | 35                                         | Aortic insufficiency, congestive heart failure              |
| Vertebral            | 35                                         | Visual changes, dizziness                                   |
| Coeliac axis*        | 18                                         | Abdominal pain, nausea, vomiting                            |
| Superior mesenteric* | 18                                         | Abdominal pain, nausea, vomiting                            |
| Iliac                | 17                                         | Leg claudication                                            |
| Pulmonary            | 10-40                                      | Atypical chest pain, dyspnea                                |
| Coronary             | <10                                        | Chest pain, myocardial infarction                           |



Vessels affected :

- Aortic arch
- Carotids
- Vertebral
- Renal.

Dx:

- Steroids - 40-60mg/day.
- Vascular surgery → treatment of stenosis
- Methotrexate.
- Tocilizumab.

Takayasu

- Medium + large vessels
- Granulomas (+)
- Skip lesions (+)
- Constitutional symptoms (+)
- Age <40 yrs
- F:M :- 8:1
- PA → PAH  
Renal.A → HTN, ↑Cr

Temporal arteritis

Medium + large vessels

- (+)
- (+)
- (+)
- Age >50 yrs
- F:M = 2-3:1
- No Pulm.A
- Renal.A involvement

Takayasu

+

-

-

ESR ↑

HLA B<sub>52</sub>Temporal arteritis

Headache + + +

Polymyalgia rheumatica  
+ + +AIOM +  
[blurred vision loss]

ESR ↑↑↑ [very high]

HLA DR<sub>4</sub>**Comparison of Giant cell arteritis & Takayasu's arteritis**

| Feature                                    | Giant Cell Arteritis    | Takayasu's Arteritis    |
|--------------------------------------------|-------------------------|-------------------------|
| Female-male ratio                          | 2:1                     | 8:1                     |
| Age range (yr)                             | ≥50                     | <40                     |
| Average age of onset (yr)                  | 72                      | 25                      |
| Visual loss                                | 10%-30%                 | Rare                    |
| Involvement of aorta or its major branches | 25%                     | 100%                    |
| Histopathology                             | Granulomatous arteritis | Granulomatous arteritis |
| Pulmonary artery involvement               | No                      | Occasionally            |
| Renal hypertension                         | Rare                    | Common                  |
| Claudication                               | Uncommon                | Common                  |
| Ethnic groups with highest incidence       | Scandinavians           | Asians                  |
| Corticosteroid responsive                  | Yes                     | Yes                     |
| Bruits present                             | Minority                | Majority                |
| Surgical intervention needed               | Rare                    | Common                  |

## VASCULITIS PART-2

### Polysystemic nodosa

- Necrotizing vasculitis → medium & small vessels.
- Viscera - Renal, hepatic, mesenteric, etc

### Not seen:

- ANCA
- Venules not involved
- Granulomas
- Eosinophilia
- Glomerulonephritis.
- Lung involvement\*\*
- M > F.
- Hep B association. → now ↓ to 5%. [due to vaccine]
- Hairy cell leukemia association.

Patho: Neutrophil infiltration → • Perivascular inflammation of vessel.  
 Acute: • Intimal proliferation/degeneration.  
 ↓  
 Chronic Mononuclear cells → Transmural inflammation.  
 ↓  
 Aneurysm, segmental\*



CFs:

- fever, malaise, fatigue.
- Arthralgia / myalgia
- HTN,  $\uparrow$  Ca.
- Mononeuritis multiplex
  - ↳ single nerve affected at multiple places
- Skin - Purpura, nodules, livedo reticularis
- GI - Abd pain, Nausea/vomiting, bloody stools

Biopsy:

- ↳ segmental transmural inflammation

Rx:

steroids + Cyclophosphamide



Livedo reticularis



Beaded appearance



## Small vessel vasculitis

ANCA

- pANCA - perinuclear - Anti-MPO.
- cANCA - cytoplasmic - Anti-proteinase-3

ANCA associated vasculitis → Pulm & renal syndromes

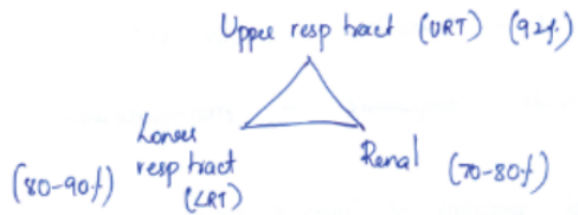
- └ Microscopic polyangiitis
- └ Wegener's [GPA]
- └ Churg Strauss [EGPA]

|                          | <u>c-ANCA</u> | <u>p-ANCA</u> |
|--------------------------|---------------|---------------|
| Wegener's                | 70%           | 25%           |
| Microscopic polyangiitis | 40%           | 50%           |
| Churg - Strauss          | 10%           | 60%           |

Wegener's (GPA)

- └ Systemic - M:F = 1:1  
Vasculitis, c-ANCA +
- └ localised - Upper resp tract - ENT  
F>M  
cANCA - ++.

## Wegener's

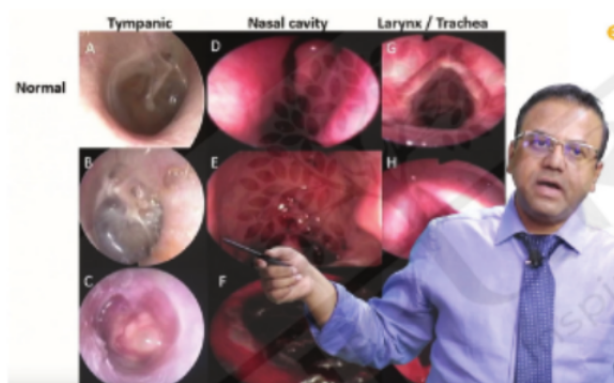
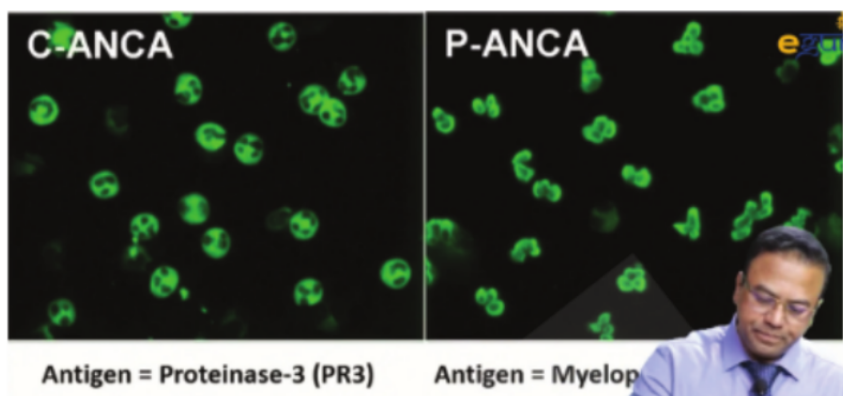


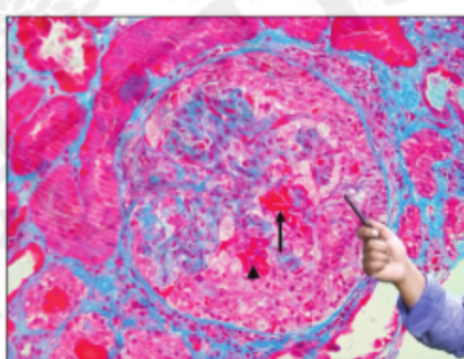
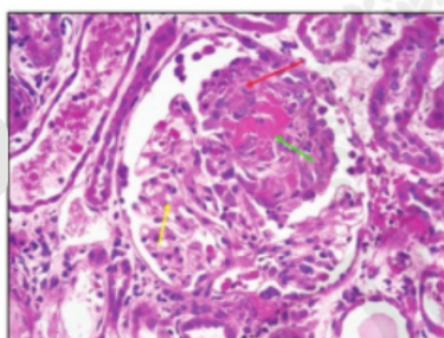
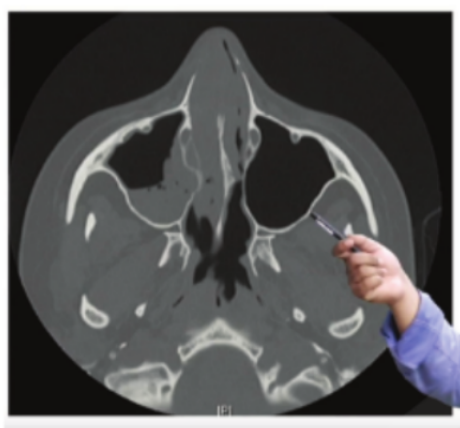
- URT → ENT - >90-95%
  - ↳ Sinusitis & headache
  - Nasal discharge
  - Otitis media → deafness. [conductive]
  - Subglottic tracheal stenosis

- LRT → Pulmonary infiltrates → Nodules
  - ↓
  - Cavitate

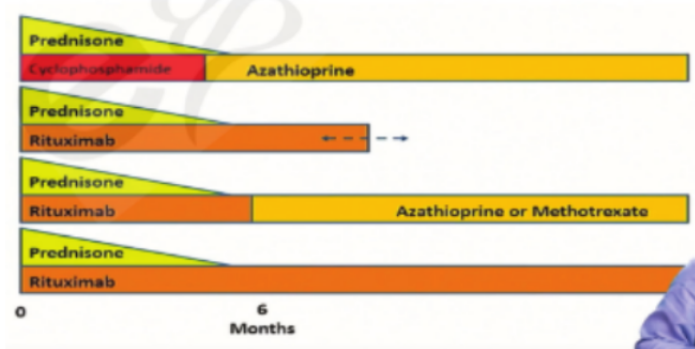
- Cough
- Chest discomfort
- Hemoptysis

- Eye → Conjunctivitis
- Episcleritis
- Scleritis





*Acute glomerulopathy.*



◦ Skin - Papules, vesicles, palpable purpura, ulcers.

◦ Renal - Focal necrotizing GN



Glomerulonephropathy.

Dr:

- Steroids + Rituximab.

- " + Cyclophosphamide.

↳ Bladder Ca.

- Methotrexate / Azathioprine. — Maintenance.

### Microscopic polyangiitis

- Renal → Pauci-immune focal necrotizing GN



Glomerulonephropathy.

- Lung → Diffuse alveolar haemorrhage.

Hemoptysis + cough.

No nodules / cavity.

- pANCA > c-ANCA

- No granulomas on biopsy

- No pulm nodules / cavities

- No/rare URT findings

### Churg - Strauss [EGPA] / AGPA (Allergic GPA)

- Prodrome → Fever, myalgia, arthralgia, fatigue
- Allergic / Asthmatic → Asthma, rhinitis, sinusitis phase
- Eosinophilic phase → Peripheral & tissue eosinophilia  
Migratory pulm infiltrates.  
Endomyocardial infiltration  
↳ Restrictive cardiomyopathy  
GI - eosinophilic infiltration.
- Vascular phase → ↓ Asthma
  - Mononeuritis multiplex\*\*
  - [Patchy sensory-motor loss]
  - Renal - Pauci immune GN
  - Skin - Purpura, ulcers

p-ANCA +

Lab:

- Eosinophilia  $>1500/\mu l$   
 $>10\%$  of total WBC.

Biopsy:

- Fibrinoid necrosis
- Eosinophilic infiltration
- Granulomas  $\oplus$ 
  - vascular
  - extravascular m.c.

GPA

URT  $\rightarrow$  sinusitis  
Nasal discharge  
Subglottic stenosis

MPA

$\ominus$

EGPA

Nasal polyps  
sinusitis

Lung:

Cavities / Nodules \*  
DAH  $+$

Diffuse alveolar  
hemorrhage (DAH)  $+$

Asthma \*  
DAH  $+$

Mononuclear infiltrates  
 $+++$

Ocular  $+++$

$\ominus$

$\ominus$

Renal - Pauci-immune necrotizing GN / Crescentic nephropathy.

Granuloma  $\oplus$

$\ominus$

$\oplus$  + eosinophilic  
infiltrates

R:

All three :

- Steroids + Cyclophosphamide
- Steroids + Rituximab
- Methotrexate, Azathioprine - maintenance

Chung- Strauss

↳ Mepolizumab  
IL-5 antagonist





## VASCULITIS - PART-3

### Henoch Schonlein purpura

- IgA vasculitis
- mc vasculitis in children
- Peak age - 4-7 yrs, can occur in adults.
- M:F → 1.5:1
- Immune complex vasculitis → small vessels.
- Palpable purpura
  - ↳ Buttocks
  - ↳ lower limbs
  - ↳ Trunk.
- [Thrombocytopenia  
↓  
Non palpable purpura]
- Arthralgia/ arthritis - large joints - knee/ankle, migratory.
- GI → abd pain, nausea, vomiting, diarrhea, blood in stools.  
 (mainly in children) Intussusception [bowel ischemia]
- Renal - Hematuria & Proteinuria  
 Biopsy → IgA nephropathy.  
 - in adults → more severe

Inv:

Serum IgA - ↑

Complement - C<sub>3</sub> - (N)

Platelet - (N)

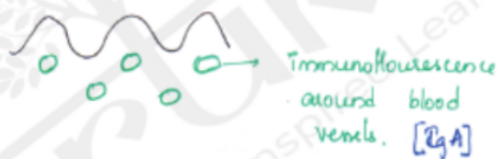
Skin biopsy - neutrophilic / monocytic infiltrates

### Diagnostic criteria:

Palpable purpura (mandatory) + at least 1 of following:

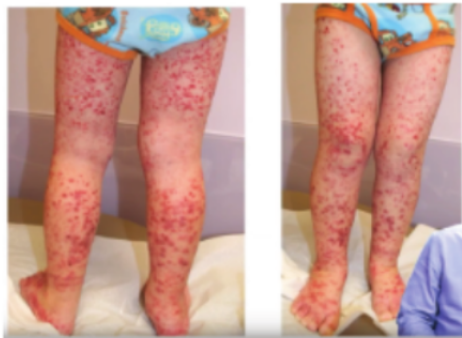
- Diffuse abdominal pain
- Arthritis (acute) or arthralgia
- Renal involvement (hematuria/proteinuria)
- Any biopsy showing predominant IgA deposition

### Immunofluorescence of skin biopsy:

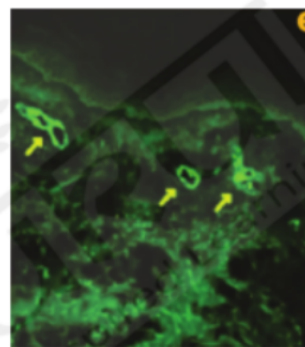


Rx:

- Steroids - 1mg/kg - good for GI symptoms joints.
- ↳ not very effective in skin, renal.



Discrete rash.



**Henoch-Schönlein purpura**

**Rash**  
 Buttocks (a)  
 Extensor surfaces of legs and arms  
 Ankles (b)

**Joint pain and swelling**  
 Knees and ankles (b)

**Abdominal pain**  
 Haematemesis and melaena  
 Intussusception

**Renal**  
 Microscopic/macroscopic haematuria (80%)  
 Nephrotic syndrome (rare)

**IgA Vasculitis**

Inflammation of glomerular capillaries

IgA deposits in glomerulus viewed by immunofluorescence microscopy

**IgA Skin Vasculitis**

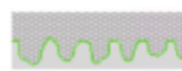
Inflammation of small vessels in the skin

IgA deposits in vessels in the skin by immunofluorescence microscopy

#### Immunofluorescence of Skin Diseases

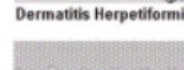


Pemphigus



Bullous Pemphigoid

#### Immunofluorescence of Skin Diseases



Dermatitis Herpetiformis



Vasculitis



Lupus Band Test

## Cryoglobulinemia

Clonal expansion of B cells  $\rightarrow$  Ig.



Precipitate on exposure to cold. [insoluble immune complex]



Block blood vessels

### Involves:

- Skin
- Nerves
- Joints
- Renal.

### Causes

Type I - **monoclonal** - lymphoproliferative causes

II - **monoclonal / polyclonal** - Hep C [Infections]  
 (M.C) ↓ Autoimmune ds lymphoproliferative  
 ↓ Rheumatoid factor (IgM) ↓ IgG

III - **polyclonal** - IgM, IgG. - Hep C, autoimmune

Essential cryoglobulinemia  $\rightarrow$  <5% - no identifiable cause.

Cls:

- Skin - Palpable purpura  
Confluent  
Ulceration
- Renal - MPGN - I [membranoproliferative GN]
- Nerves - Mononeuritis multiplex  
Neuropathy
- Joint - small joint  
Arthralgia
- Pulmonary haemorrhage → hemoptysis

Inv:

- Blood test :  
 Store blood - 5-6 days  
 ↓  
 Centrifuge → ppt (+)  
 ↓  
 dissolves on re-suspending
- Complement -  $C_4 \downarrow$

- Tx:
- Treat underlying cause
    - Rituximab
    - Steroids - cautious in HepC infection
    - Plasmapheresis



Confluent purpura



### Behcet's

- Recurrent oral/genital ulcers with ocular manifestations
- HLA B5
  - ↳ subtype B51.
- Perivasculitis involving all kinds of vessels.
- Systemic - vasculitis
  - ↳ Neutrophil infiltration
  - ↳ endothelial swelling.

$T_H1$ ,  $T_H17$ , cytotoxic  $CD8$  cells involved.
- Cfs:
  - Essential feature: Recurrent oral aphthous ulcers
    - ↳ Small <10mm
    - ↳ Single
    - ↳ last 1-2 weeks
    - ↳ painful
    - ↳ heal without scarring.

Yellow necrotic base with halo around.
  - Genital ulcers - painful
    - Do not affect glans penis, vagina, cervix
    - Involves scrotum, vulva
    - heal with scarring.

- Oculae - Anterior segment / posterior segment.
  - Uveitis - Panuveitis → **deadly**.
  - Retinal vasculitis
  - Optic neuritis
  - Hypopyon → **aggressive disease & poor prognosis**

- Skin -
  - Erythema nodosum
    - ↳ involve lower limbs
  - Pyoderma gangrenosum
  - Acne like eruptions.

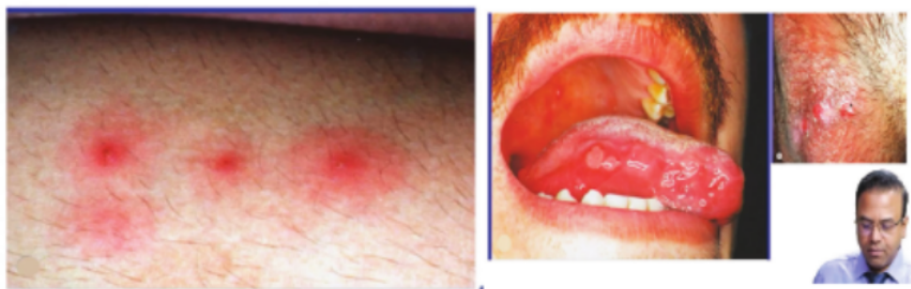
Inv:

**Pathergy test:** Non specific inflammatory response to saline injection in skin  
- interpreted at 24-48 hrs.

- Venous thrombosis: Cortical vein thrombosis
- Superficial thrombophlebitis
- Neuro (10+) - Parenchymal - brainstem, pyramidal, cerebellum

**CSF → ↑ IL-6**





Pathergy test

Oral &amp; genital ulcers



Erythema nodosum

Hypopyon &amp; Panuveitis

- GI - ulcers
- Amyloidosis
- Epididymitis

### Diagnostic criteria :

Recurrent oral ulceration + 2 of following :

- Recurrent genital ulceration
- Ocular lesions
- Skin lesions
- Positive patchy test.

Rx :

(POC) → Colchicine.

Ocular - Azathioprine  
TNF inhibitors  
Calcineurin inhibitors  
Steroids

Mucocutaneous - Topical steroids  
(aggressive)

Vascular - Steroids + Cyclophosphamide / Azathioprine

CNS - Cyclophosphamide / Aza + Steroids

## Relapsing polychondritis

Autoimmune disease targeting cartilage



Ears / Nose / Eye / Trachea

CGs:

- U/L - B/L - auricular chondritis  
[does not involve ear lobe]
- Nasal cartilage → septal perforation
  - Saddle nose deformity
- Conductive / sensorineural hearing loss
- Symptoms → days / weeks → Resolve.
- Laryngeal / tracheal cartilage → airway obstruction
  - ↳ Cough, throat soreness, hoarseness, pain in tracheal / thyroid cartilage.
  - ↳ Glottic strictures.
- Non erosive arthritis
- Aortitis

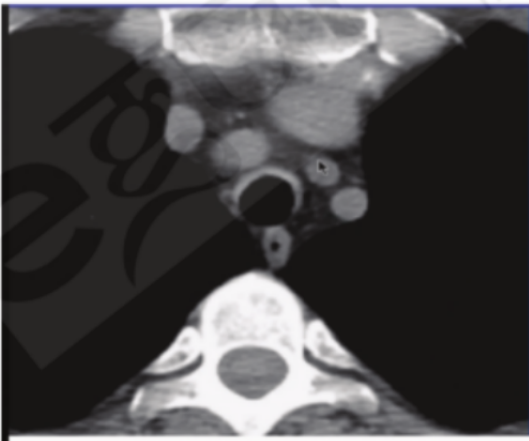
Diagnosis:

- mainly clinical
- Cartilage biopsy
- Pulm function test
- **Dynamic CT**

↳ difference in tracheal lumen in insp & exp.  
[tracheal diameter]

Tx:

- Mild - NSAIDs  
Low dose steroids
- Severe - High dose steroids



### Kawasaki disease

- exclusively seen in children
- Anti- endothelial Abs
  - ↳ medium sized vessels
  - ↓
  - Coronary A. aneurysms +
- Fever  $\times$  5 days - essential criteria.
  - +  $\geq 4$  of following:
    - Cervical lymphadenopathy  $\rightarrow$   $>1.5$  cm
    - Conjunctivitis
    - Polymorphic rash - trunk
    - Mucosal - Strawberry tongue, dry cracked lips
    - Peripheral - UL extremities  $\left\{ \begin{array}{l} \text{Swelling} \\ \text{Desquamation} \end{array} \right.$   
 LL

Tx:

Aspirin + IV. IgG.

**Table 1. Diagnostic criteria for Kawasaki disease**

Diagnosis requires unexplained fever for  $\geq 5$  days in addition to the presence of  $\geq 4$  of the following:

- Oral mucosa changes, including red or cracked lips, pharyngeal erythema, or strawberry tongue
- Bilateral nonexudative conjunctivitis, sparing the limbus
- Cervical lymphadenopathy, usually unilateral, with one node  $\geq 1.5$  cm in size
- Polymorphous rash
- Extremity changes (erythema of palms and soles, swelling of hands and feet, periungual desquamation in the convalescent phase)

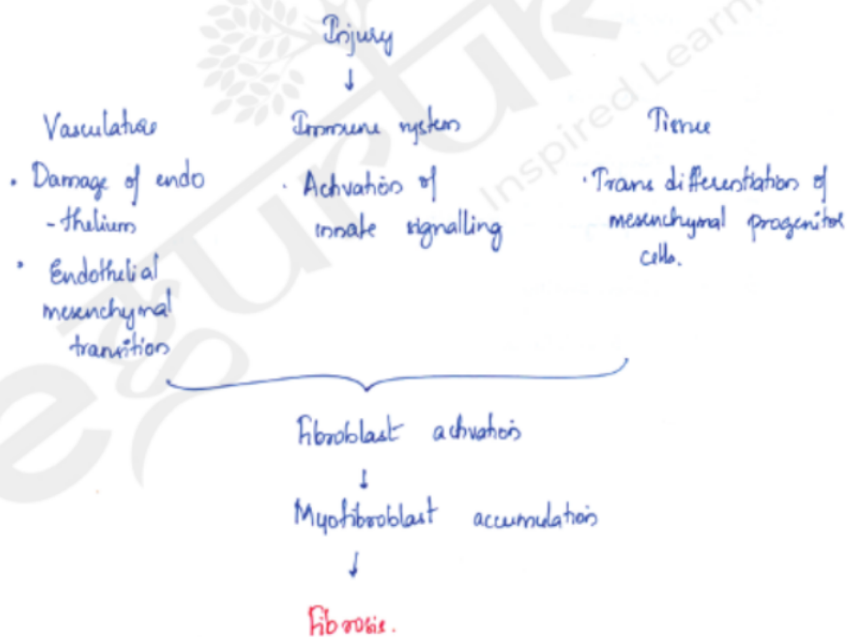


## Scleroderma systemic sclerosis Part-1

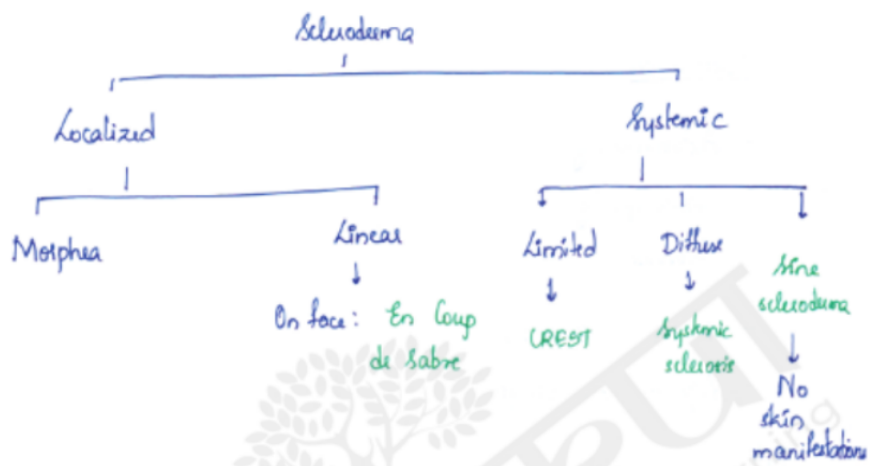
### Scleroderma

- Autoimmunity
- Vasculopathy
- Fibrosis
- F:M = 3-4:1
- Age - 30-60 yrs

Patho:



Classification:



CREST syndrome:

- C - Calcinosis
- R - Raynaud's
- E - Esophageal dysmotility
- S - Sclerodactyly
- T - Telangiectasia



### Systemic sclerosis :

- S - Skin sclerosis
- C - Cardiac
- L - Lung fibrosis
- E - Esophageal dysfunction
- R - Raynaud's
- O - Obstruction - bowel
- D - Digital ulcers
- E - Endocrine (Thyroid - Hypo)
- R - Renal crisis
- M - Myositis
- A - Arthralgia / Arthritis

| <u>Limited ss</u>                   |                                                                                     | <u>Diffuse ss</u>                               |
|-------------------------------------|-------------------------------------------------------------------------------------|-------------------------------------------------|
| +                                   | Calcinosis                                                                          | +                                               |
| Early - +                           | Raynaud's                                                                           | + along with skin inv.                          |
| +                                   | Eso dysmotility                                                                     | +                                               |
| +                                   | Sclerodactyly                                                                       | +                                               |
| +                                   | Telangiectasia                                                                      | +                                               |
|                                     | (Peknungal)                                                                         |                                                 |
| Raynaud's precedes skin involvement |  | Raynaud's & skin manifestations simultaneously. |

Limited SS

- Anticentromere Ab
- Th/To Abs

Diffuse SS

- Anti scl-70 Abs (topoisomerase)
- RNA polymerase-3 Abs

## Skin fibrosis:

Below elbow  
Below knee  
Above neck

Above elbow  
Above knee  
Below neck / trunk\*

## Lung:

Rare\*

Interstitial lung ds (70%)

Pulm HTN ⊕ - Primary  
Pulm HTN.

⊕

## Renal crisis:

2%

Crises - 15%

| TABLE 353-3 Subsets of Systemic Sclerosis (SSc): Features of Limited Cutaneous SSc versus Diffuse Cutaneous Disease |                                                                                                        |                                                                                                     |
|---------------------------------------------------------------------------------------------------------------------|--------------------------------------------------------------------------------------------------------|-----------------------------------------------------------------------------------------------------|
| CHARACTERISTIC FEATURE                                                                                              | LIMITED CUTANEOUS SSc                                                                                  | DIFFUSE CUTANEOUS SSc                                                                               |
| Skin involvement                                                                                                    | Indolent onset. Limited to fingers, distal to elbows, face; slow progression                           | Rapid onset. Diffuse: fingers, extremities, face, trunk; rapid progression                          |
| Raynaud's phenomenon                                                                                                | Antedates skin involvement, sometimes by years; may be associated with critical ischemia in the digits | Onset coincident with skin involvement; critical ischemia less common                               |
| Musculoskeletal                                                                                                     | Mild arthralgia                                                                                        | Severe arthralgia, carpal tunnel syndrome, tendon friction rubs; small and large joint contractures |
| Interstitial lung disease                                                                                           | Slowly progressive, generally mild                                                                     | Frequent, early onset and progression, can be severe                                                |
| Pulmonary arterial hypertension                                                                                     | Frequent, late, may occur as an isolated complication                                                  | Often occurs in association with interstitial lung disease                                          |
| Scleroderma renal crisis                                                                                            | Very rare                                                                                              | Occurs in 15%; onset may be fulminant; generally early (<4 years from disease onset)                |
| Calcinosis cutis                                                                                                    | Frequent, prominent                                                                                    | Less common, mild                                                                                   |
| Characteristic autoantibodies                                                                                       | Anti-centromere                                                                                        | Anti-topoisomerase I (Scl-70), anti-RNA polymerase III                                              |

### Skin :

- Puffiness - fingers / edema
- Skin thickening
- Hyper / hypo pigmentation
- Pinched nose
- ↓ oral aperture
- Scleroderma 'neck' sign.
- **Mask facies**
- **Modified Rodnan skin score** - 17 areas

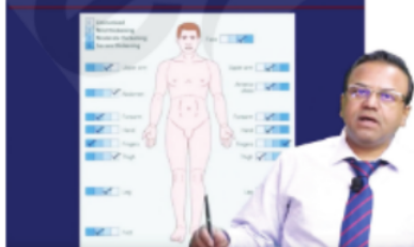
Puffy hands



Mauskopf Facies



Modified Rodnan Skin Score



## Scleroderma systemic sclerosis Part - 2

Raynaud's phenomenon:

↳ >90% of pt with systemic sclerosis.

↑ vasoconstriction → Raynaud's phenomenon.

### Primary Raynaud's

- No association
- B/L symmetric
- Reversible.
- Rare - ulcers/gangrene
- <40 yrs

### Secondary Raynaud's

- Associated with connective tissue disorders, drugs, vascular.
- Asymmetric
- Ulcers/gangrene/Acral osteolysis - common
- >40 yrs

Rx:

- Avoid exposure to cold
- CCBs - Nifedipine - sustain release  
Amlodipine
- PDE-5 inh → Sildenafil, Tadalafil - Add. to CCBs
- Nitroglycerine - topical.
- Endothelin receptor blocker - Bosentan.

- i.v prostacyclin analogue - Epoprost.
- Sympathectomy.

Patient w/ Raynaud's.

↓  
to differentiate Primary or CREST.

↓  
Nail fold capillaroscopy. / ophthalmoscope.

Normal

- Open hairpin like
- Symmetrically arranged
- 7-14



↓  
Primary Raynaud's

• Large capillaries

• Telangiectasia

↓↓ → later capillary loops ↓.

↓  
CREST syndrome



GIT

- 80-90% affected
- Esophageal motility → Dysphagia  
GERD  
Barrett's metaplasia
- Smooth muscle atrophy + esophageal wall fibrosis
- Dilated esophagus → ↓ motility  
↓ LES tone.
- Gastric antral venous ectasia (GAVE)
  - ↳ Fe def anemia
  - ↳ Chronic blood loss
  - ↳ Endoscopy: Watermelon stomach.
  - Ⓡ: laser photocoagulation.
- Small bowel:
  - ↑ transit time → Ⓡ - Erythromycin, metoclopramide.
  - Diverticulosis
    - ↳ bacterial overgrowth syndrome
    - ↓
    - Malabsorption
    - Ⓡ: Hydrogen breath test.
    - Ⓡ: Rifaximin.

- Anal sphincter laxity → Fecal incontinence.

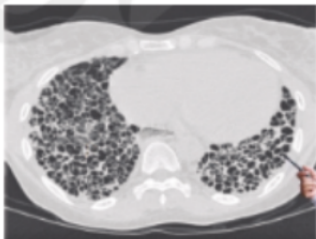
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- Interstitial lung fibrosis (ILD) → HRCT. - lower lobes
- Pulm HTN
  - due to ILD
  - primary (vasculopathy)
- Cancer
- Pleural effusion
- Musculoskeletal → ↓ chest expansion → Dyspnea.
- Aspiration
- Infection
- Airway disease.

### Biopsy:

- Non-specific interstitial pneumonitis

Follow-up is by pulmonary function tests (PFTs)



| <u>HRCT</u>   | <u>Treatment</u>               |
|---------------|--------------------------------|
| <20%          | No treatment                   |
| >20%          | Immunosuppression              |
| Indeterminate | Use FVC to determine treatment |
|               | <70% : Treat                   |
|               | >70% : Don't treat             |

Re:

Myophenolate

Rituximab / Cyclophosphamide / Azathioprine

Nintedanib → GI intolerance

↳ intracellular tyrosine kinase inhibitor

Low dose steroids → Renal crisis

Pulm HTN:

mean Pulm artery pressure > 25 mmHg.

Re:

- O<sub>2</sub>

- Bosentan

- PDEI

- Prostacyclin analogues



Cardiac:

- Diastolic dysfunction
- Conduction blocks
- Pericardial disease - Pericardial effusion

Renal crisis

- Hypertensive spikes
- Microangiopathic hemolytic anemia (MAHA)
  - ↳ Schistocytes
- Fundus → HTN retinopathy
- ↑ Cr.

Rx: ACEI.

↓  
Captopril - recommended.

↑ risk of renal crisis:

Steroids

RNA polymerase-III Abs

First 3 years

Diffuse systemic sclerosis

Tendon friction rubs.

### Immunofluorescence :

Speckled - Scl-70  
 Nucleolar - RNA pol III  
 Th/To  
 PM - Scl  
 Centromere - Centromere - Specific for  
 CREST syndrome.

| <u>Ab</u>               | <u>Risk</u>                                     |
|-------------------------|-------------------------------------------------|
| Anti-centromere         | Associated with PAH                             |
| Anti scl-70             | Associated with progressive ILD                 |
| Anti RNA polymerase III | Renal crisis & malignancy                       |
| Anti PM - Scl           | Overlap disorder associated with ILD & myositis |

### Re of systemic sclerosis

|                                                         |                                                            |
|---------------------------------------------------------|------------------------------------------------------------|
| Raynaud's<br>GI dysmotility<br>Pulm HTN<br>Renal crisis | } Symptomatic treatment.<br>[No need of immunosuppression] |
|---------------------------------------------------------|------------------------------------------------------------|

- Diffuse skin involvement
  - Progressive ILD
  - Inflammatory arthritis
  - Myositis
- } Immunosuppression.

Skin  
Progressive ILD } Mycophenolate  
Inflamm arthritis - Hydroxychloroquine  
Myositis - low dose steroids.

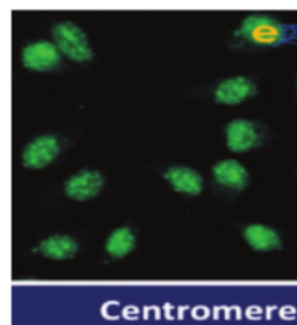
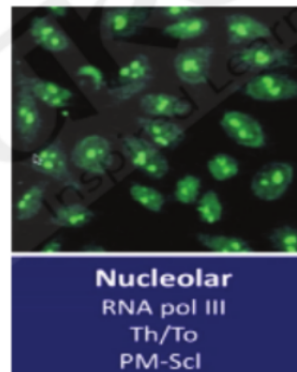
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Skin manifestations like scleroderma without Raynaud's, without Abs.

- Scleroderma : **Diabetes**  
Woody induration of skin → back.
- Scleromyxedema : waxy feel with papules
- Eosinophilic fascitis : Eosinophilia  
Grooves in skin [Empty river bed sign]  
Hands not involved  
No Raynaud's phenomenon.  
Rx: Steroids

- Nephrogenic systemic fibrosis:
  - ↳ Gadolinium used in patients with  $GFR < 30$ .
  - Thickening of skin - extremities & trunk.
  - CD34+ fibrocytes.

Eosinophilic fascitis



## SYSTEMIC LUPUS ERYTHEMATOSIS - PART 1

- F:M = 9:1
- 20-60 years



- Genes :
- TREX1 gene
  - Klinefelter's syndrome. [more the X chr → ↑ risk]
  - Complement deficiency  
[Complement help in clearing apoptotic bodies]
  - HLA - DRB1, DR3, DQA2
  - APOE1 (African americans)
  - STAT4

- Environment :
- UV light
  - Smoking
  - Crystalline silica
  - EBV infections

Cfs:

Mnemonic : MD SOAP BRAIN

- Malar rash - Butterfly rash on face  
sparing nasolabial folds
- Discoid rash - heals with scars
- Serositis - Pleuritis - Pl effusion  
Pericarditis - Pericardial effusion  
Ascitis
- Oral ulcers - > soft painless  
Hard palate.
- Arthritis - Small joints  
Non-erosive
- Photosensitivity - UV-B
- Blood dyscrasias - Anemia  
Thrombocytopenia  
Leukopenia
- Renal abn -
- Abs - ANA
- Immunological

- Neurological - Organic brain syndrome  
Behavioral abn<sup>o</sup>., Cognitive dysfunction.  
Seizures  
Psychosis

Skin - 90% of patients

Acute : Malaise rash, erythema, interarticular rash

Subacute : Annular, Psoriasisiform

Chronic : Discoid rash

↳ Mnemonic : PASTE.

Plugging

Atrophy

Scarring

Telangiectasia

Erythema

- Can occur even without SLE [Abs ⊖]

Oral ulcers :

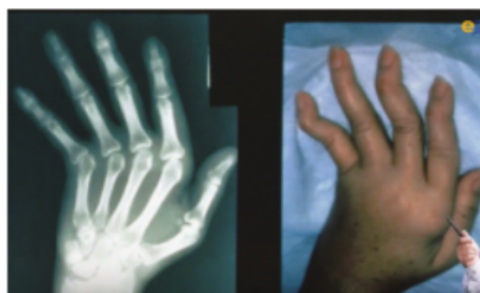
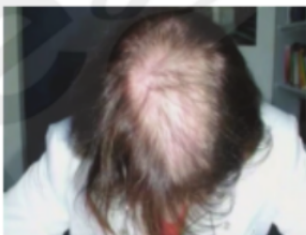
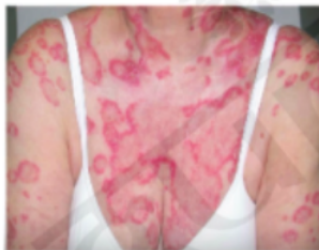
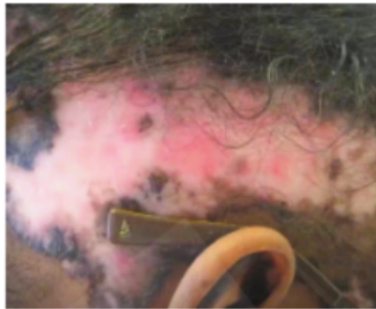
- Palate, Tongue, buccal mucosa, nasal
- Painless mostly.

- Alopecia - Non-scarring alopecia.
- Synovitis - Arthralgia,  
Arthritis - Symmetric polyarthritis  
Small joints - MCP.  
Non-erosive\*

**Jaccoud's deformity** - ulnar deviation of fingers  
- Correctable.







SLE      Part 2

Leucositis : Pleuritis → chest pain worsening on coughing

Pl. effusion

Pericarditis

Pericardial effusion

Peritonitis

Leucitis

Renal : 70f

Minimal - Non nephrotic proteinuria  
Hematuria

Nephritis - HTN, ↑G, Edema

Nephrosis - Proteinuria (Nephrotic), Edema, Thrombosis

ISN classification: [Patho - biopsy based]

Class I - Minimal mesangial

2 - Mesangio proliferative

① - Focal nephritis (<50% Glomeruli)

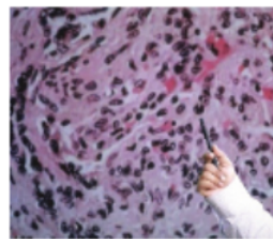
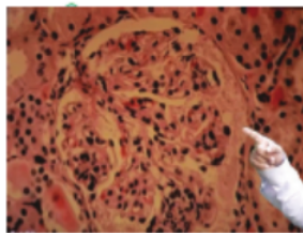
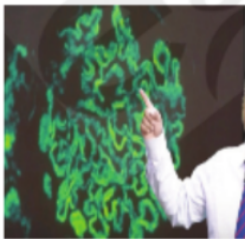
- Active - Crescents (+), subendothelial deposits  
[wire loop lesions]  
fibrinoid necrosis
- Chronic - Fibrous crescents, adhesions, sclerosis

- IV - (>50% of glomeruli involved)  
 Diffuse nephritis  
   ↳ Active - Segmental  
   ↳ Chronic - Global
- V - Membranous
- VI - Advanced sclerotic nephrotic

#### Biopsy in SLE :

- Unexplained ↑Cr
- Proteinuria >1g
- Proteinuria >500mg + Hematuria
- Active urine sediment.

- Class I, II - Don't treat
- III, IV, V - Treat
- VI - Don't treat.



Pulm :

- Pleuritic, Pl. effusion → mc
- Parenchyma - Non specific interstitial pneumonitis (rare)
- Acute lupus pneumonitis - Cough, chest pain, Hypoxemia, fever, Xray - pulm infiltrates.
- Diffuse alveolar hemorrhage.
  - ↳ Hemoptysis, dyspnea, ↓ SaO<sub>2</sub>, ↑ D<sub>L</sub>CO.
- Shrinking lung syndrome.
  - ↳ Restrictive pattern.

Cardiac :

- Asymptomatic pericarditis
  - ↓
  - Asymptomatic → Chest pain
- Coronary artery disease → 2-10 fold ↑.

Neurological :

- Neuropsychiatric
- Headache, cognitive decline, mood disorder
- Seizures
- Behavioral abn
- Myelitis, peripheral neuropathy.

Blood :

- Anemia
- Neutropenia
- Thrombocytopenia

2012 - Systemic lupus international collaboration criteria

- Skin
  - Acute
  - Subacute
  - Chronic
- Alopecia
- Oral ulcers
- Arthritis
- Renal abn
- Blood abn
- Neurological

Lab :

- ANA
- Anti ds DNA
- Anti - Sm
- Antiphospholipid
- Coombs test
- Complement

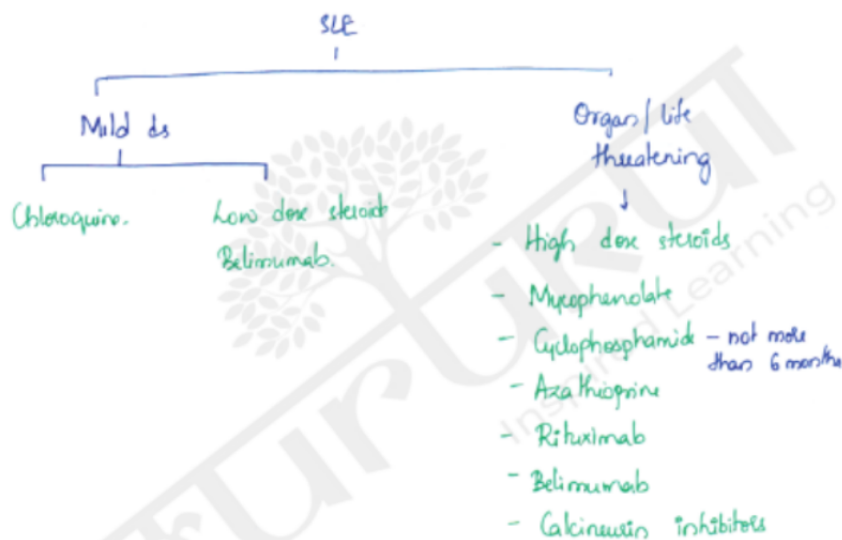
Diagnosis:

Total 4 ⊕ → 1 at least from clinical & 1 from lab.

Kidney biopsy ⊕ → Diagnosis - no need of criteria.

R:

- Hydroxychloroquine - most important drug.
- Supportive Rx
- Steroids



- Mycophenolate → Inosine monophosphate dehydrogenase inhibitor
- Belimumab → monoclonal Ab against BAF
- ↓ steroid dose.

### SLE & Pregnancy

- Renal & CNS well controlled for 6 months
- - Hydroxychloroquine
  - Low dose steroids } continued
- If Anti-SS(a) → check fetus for conduction abn
- Antiphospholipid Abs (+) → LMWH,  
Low dose aspirin

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↳ >90% of pt with systemic sclerosis.

↑ vasoconstriction → Raynaud's phenomenon.

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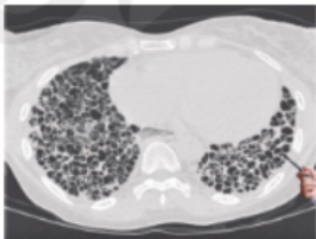
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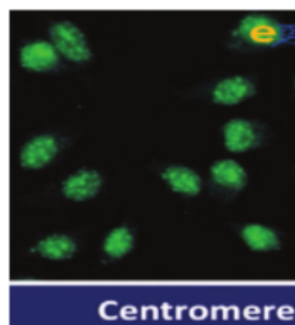
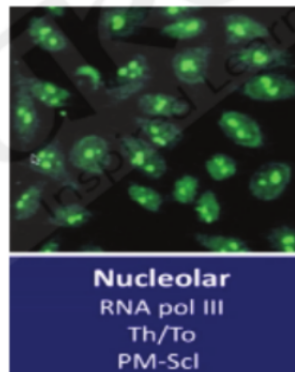
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